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MEDICAL

Neurology

Dr. Conrad Fischer

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Chapter 1: Diseases of the Spinal Cord

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Spinal Cord Compression

Spinal Cord Compression — Etiology

- This is a neurologic emergency!
- Secondary to
  - Cancer
  - Epidural abscess
  - Hematoma

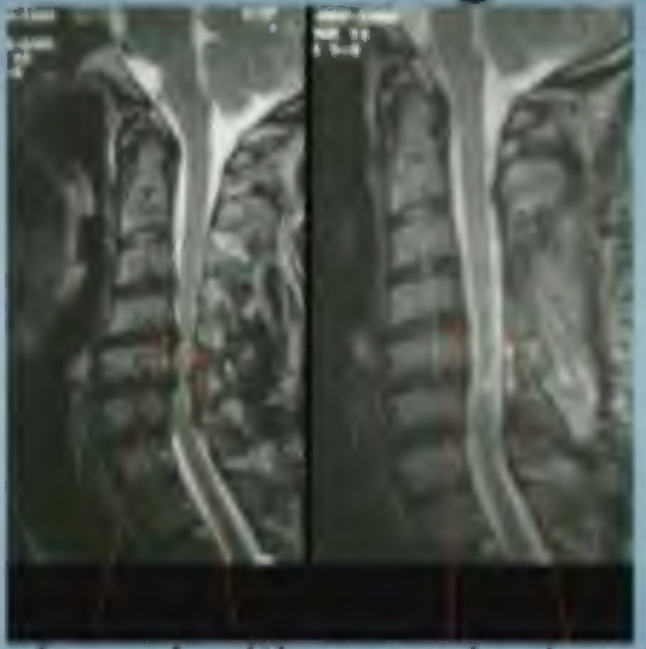
Spinal Cord Compression — Presentation

- Insidious
  - Sensory disturbance
  - Lower extremity weakness
  - Sphincter dysfunction
  - Pain
  - Hyperreflexia

Spinal Cord Compression — Treatment

- High dose dexamethasone
- Further treatment differs with etiology
  - Tumors
    - Radiotherapy
  - Herniation, abscess, or hematoma
    - Surgical decompression

Spinal Cord Compression: Before and After Neurosurgical Repair



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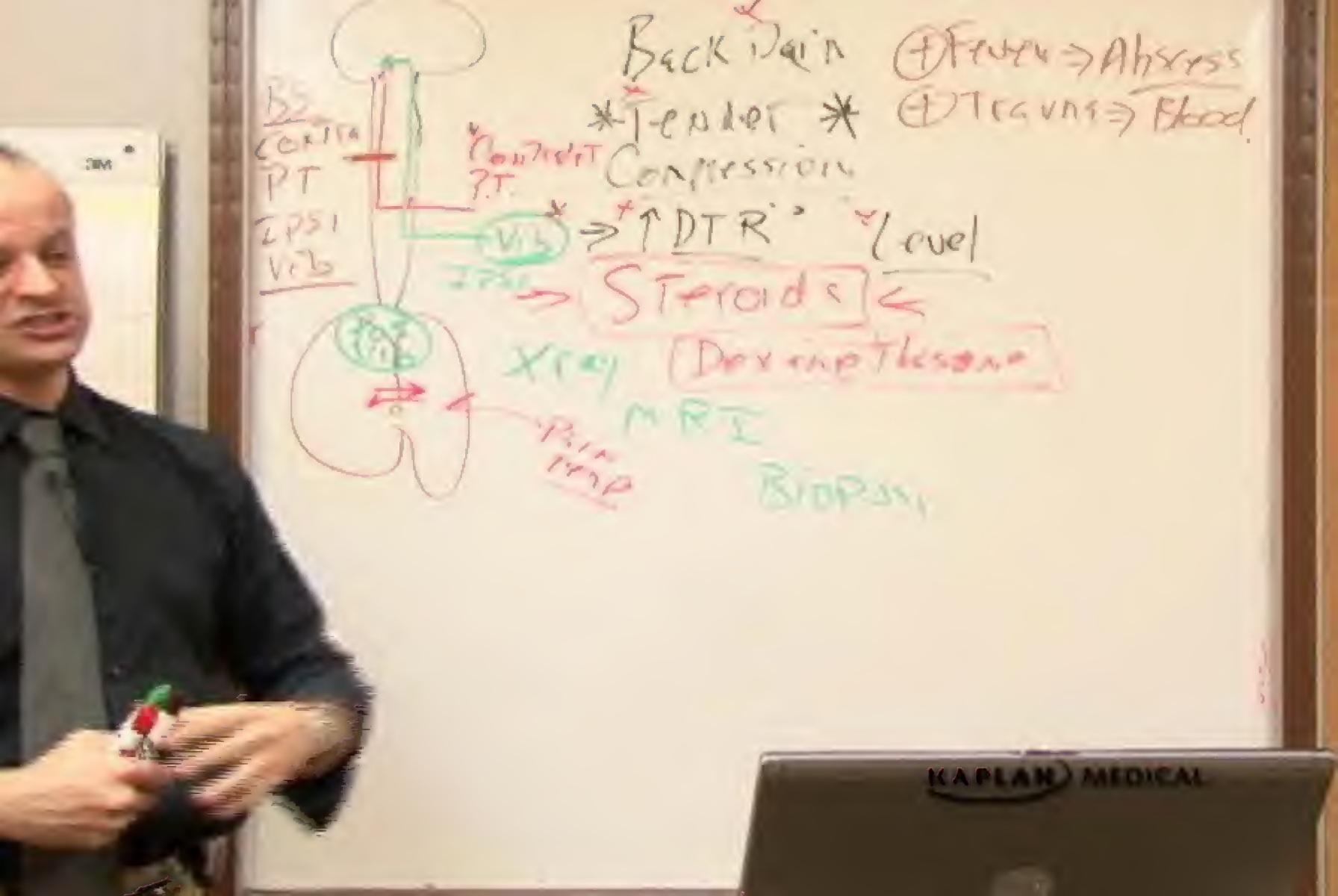
Back Pain
(+) Fever ⇒ Abscess
(+) Trauma ⇒ Blood
* Tender *
Compression
⇒ ↑ DTR
Level
Steroids
Dexamethasone
MRI
Biopsy
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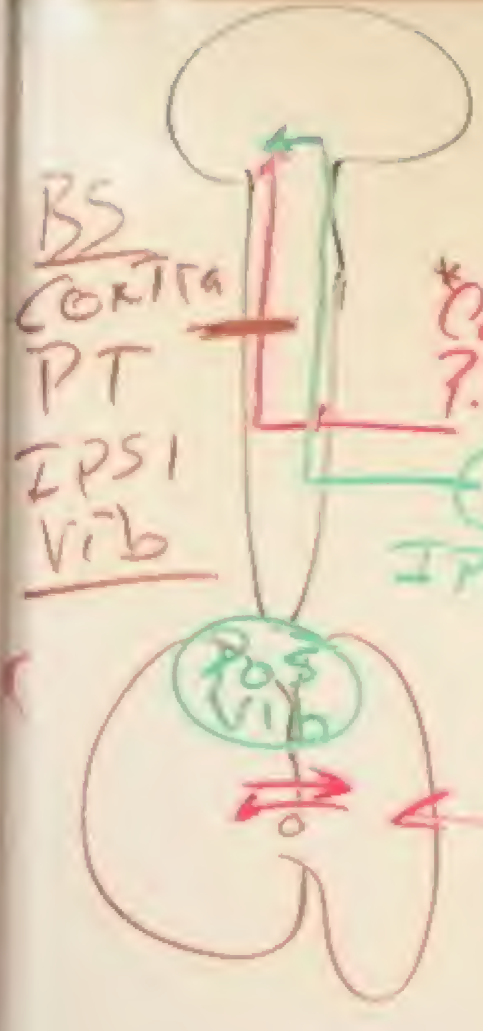

Back Pain

(+) Fever ⇒

(+) Trauma

* Tender *

Compression

⇒ ↑ DTR's

Level

Steroids

X-ray

MRI

Biopsy

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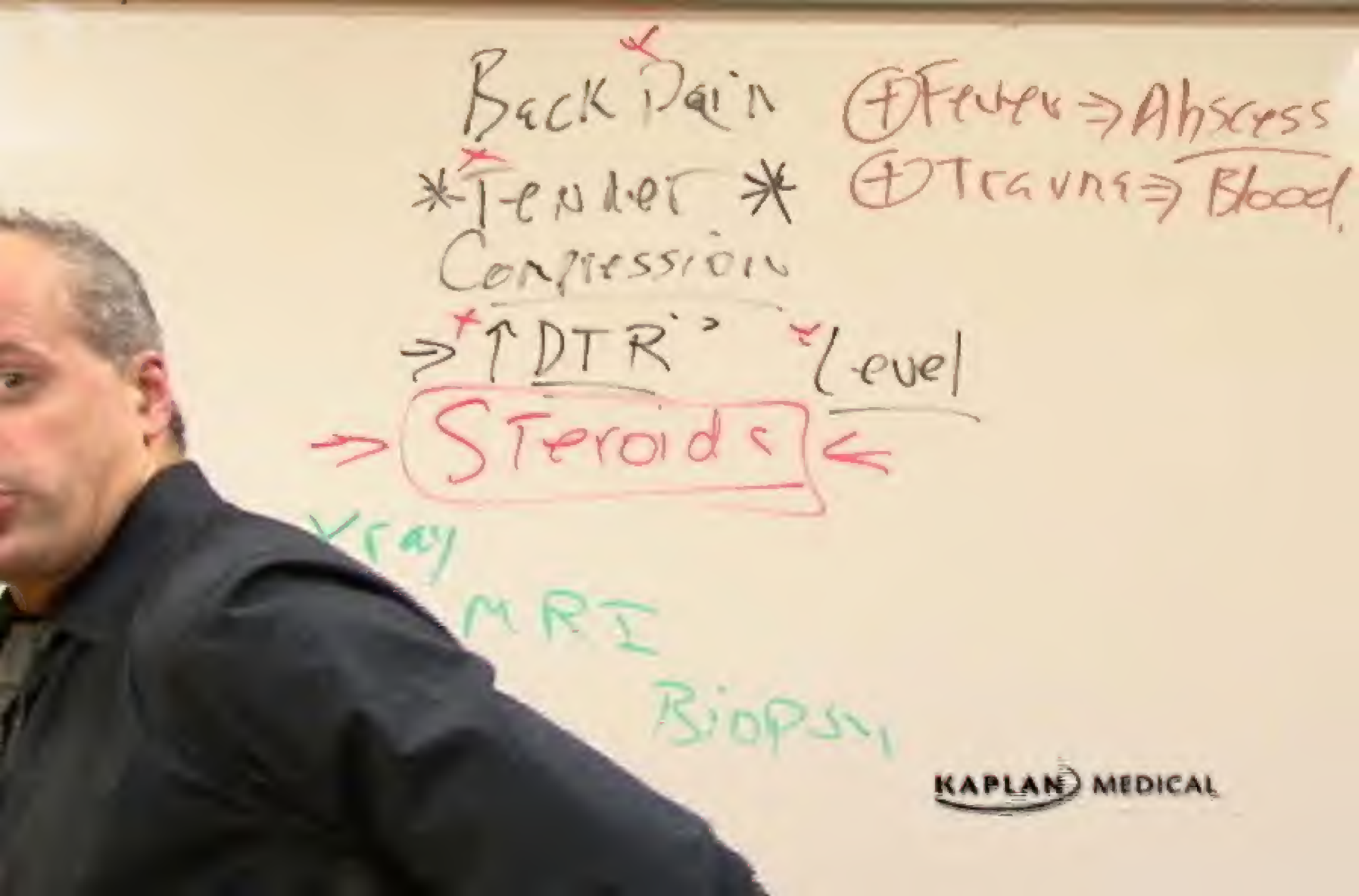
Back Pain
Tender
Compression
⇒ ↑ DTR's
Level
⇒ Steroids ⇐
X-ray
MRI
Biopsy
(+) Fever ⇒ Abscess
(+) Trauma ⇒ Blood
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Spinal Cord Compression

END

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Syringomyelia

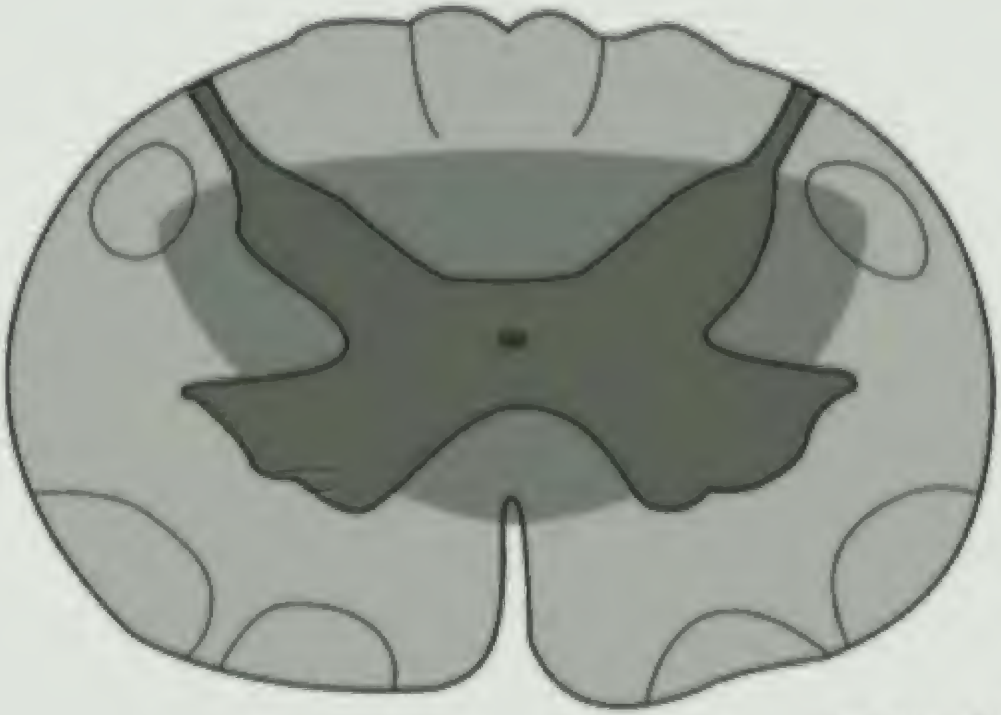
Syringomyelia — Etiology

- Most common in the cervical spinal cord
- Communicating
  - Arnold-Chiari Malformation
- Non-Communicating
  - Trauma
  - Tumors

Syringomyelia — Presentation

- Cape-like loss of pain, temperature and sensation to light touch across the neck and arms
- Sparing of tactile sensation, position and vibration
- Absent reflexes

Syringomyelia



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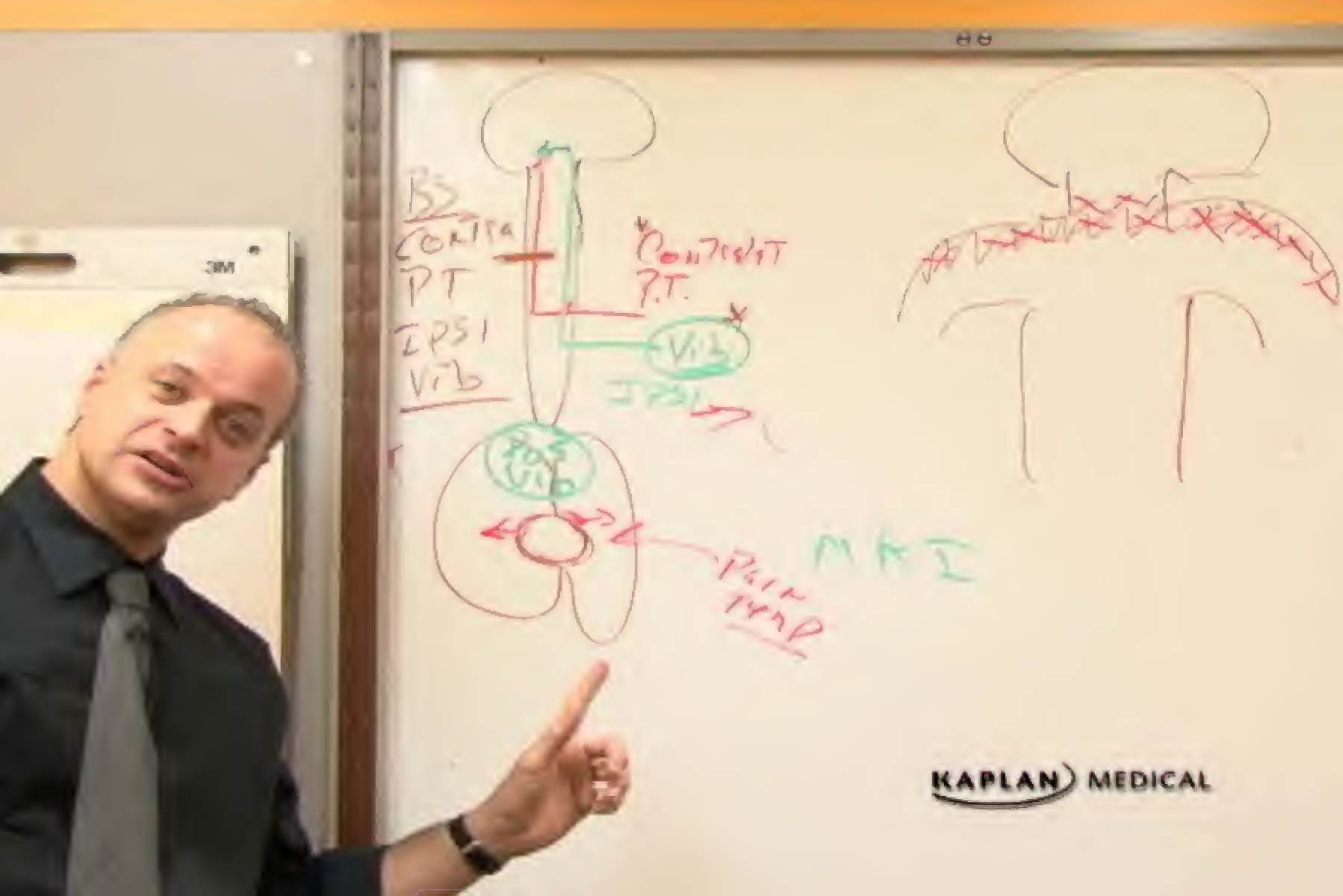
Syringomyelia — Treatment

- Surgical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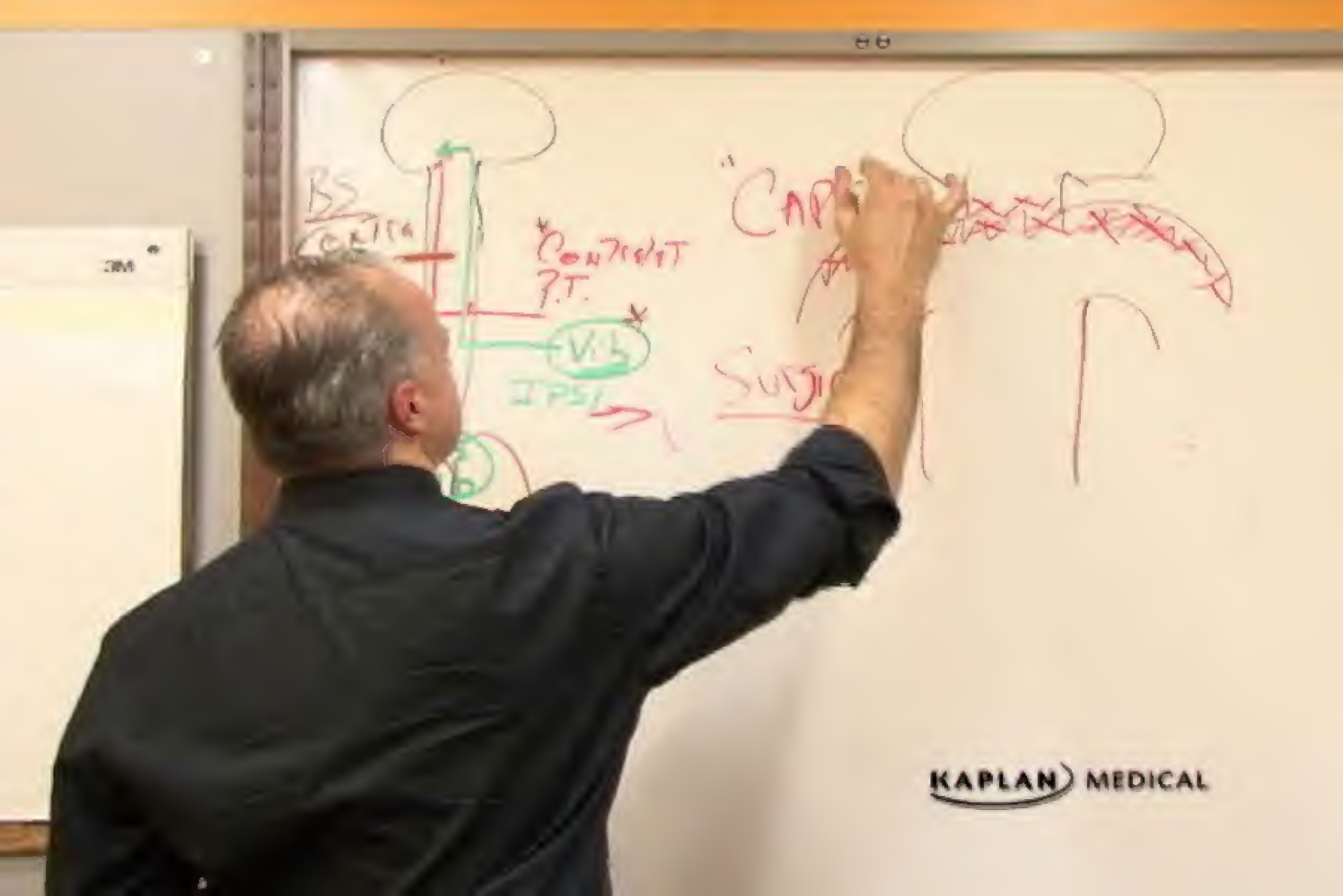
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Syringomyelia

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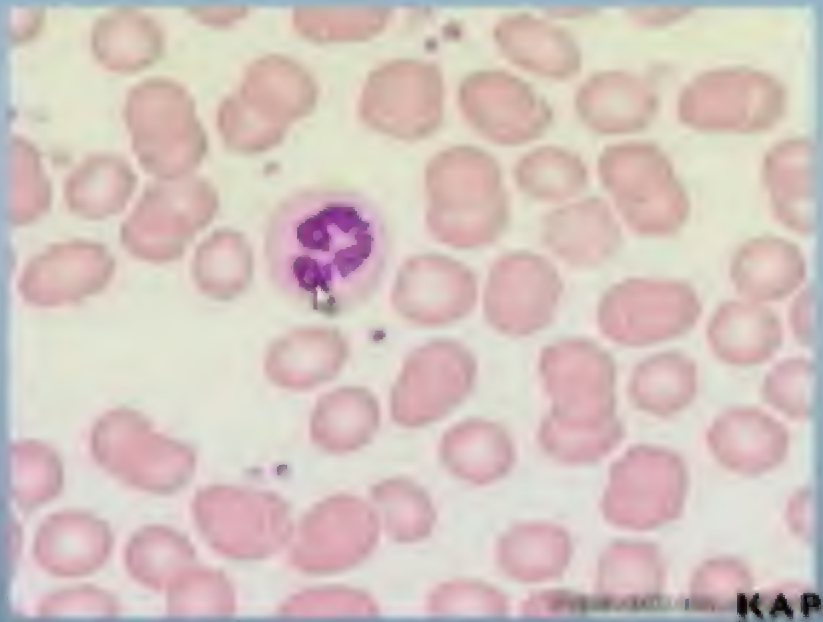
Subacute Combined Degeneration

Subacute Combined Degeneration — Etiology

- Secondary to B-12 deficiency

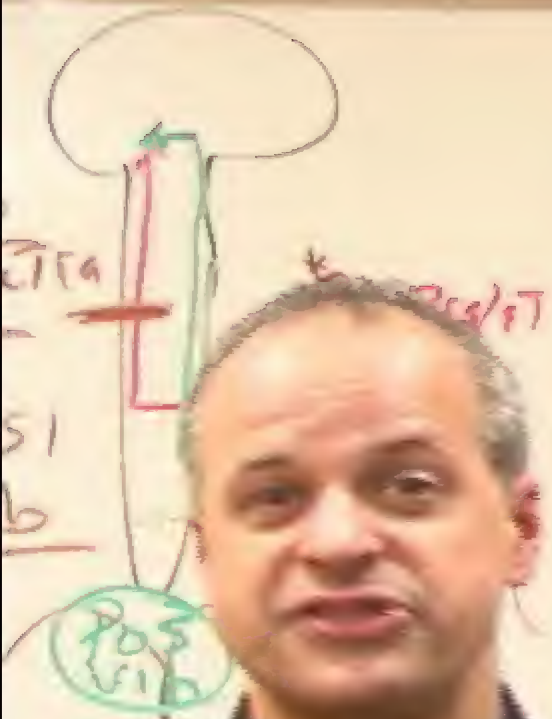


Subacute Combined Degeneration — Clinical Presentation

- History:
  - Distal paresthesias and extremity weakness
  - Progresses to spastic paresis and ataxia if left untreated
- Physical Exam
  - Combined deficit of vibration and position sense
  - Pyramidal signs

Subacute Combined Degeneration — Diagnosis and Treatment

- Diagnosis
  - Low serum B-12
  - May have macrocytic anemia with hypersegmented neutrophils
- Treatment
  - B-12 replacement



Neuro
*Peripheral

Macro ⇒ ↑MCV B12.
Ovalocytes
↑Bili ↑LDH

Megaloblastic B12/Folate
↑ Segments

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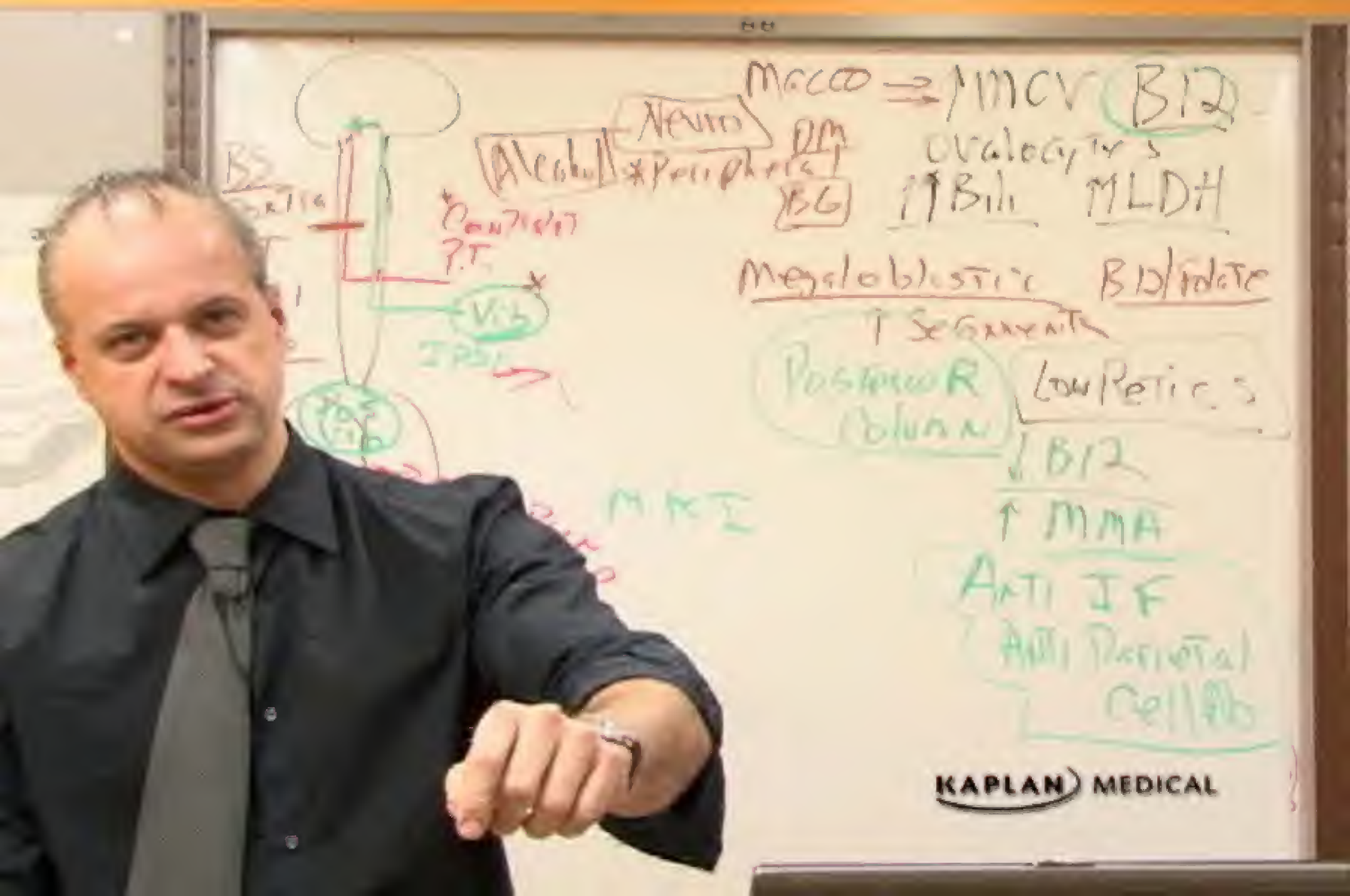
MCC ⇒ MCV B12
Neuro
Alcohol
Peripheral
Megaloblastic
Posterior Column
Low Retics
↓ B12
↑ MMA
Anti IF
Anti Parietal Cell Ab
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B12 Malabs

↑ Segments

Posterior Column

Low Retics

↓ B12

↑ MMA

Anti IF

Anti Parietal Cell Ab

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Subacute Combined Dengeneration

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Anterior Spinal Artery Occlusion

Anterior Spinal Artery Infarction — Presentation

Acute onset of flaccid paralysis

Days to weeks later

Spastic paresis

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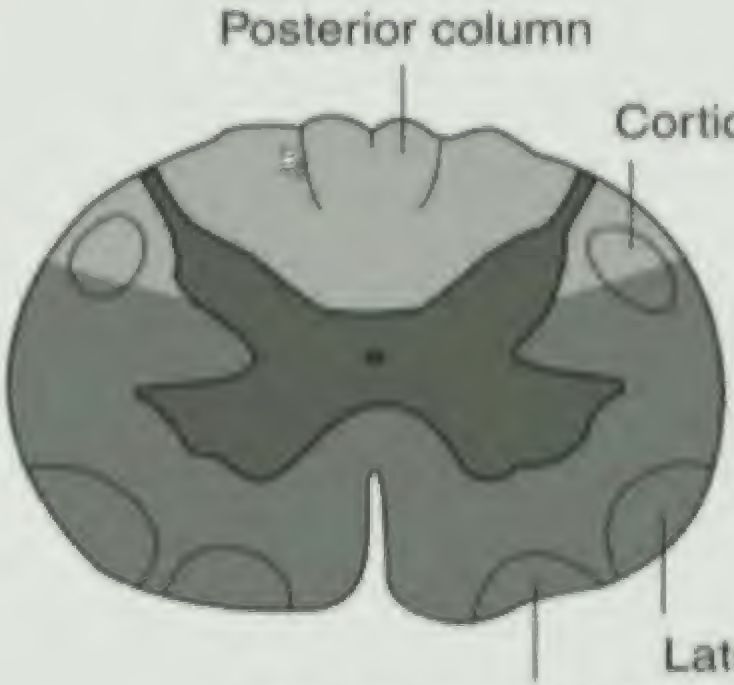
Anterior Spinal Artery Infarction — Presentation

- Everything is lost except position and vibratory senses
  - Pain and temperature- Lateral spinothalamic tracts
  - Motor- Anterior columns
  - Autonomic- Intermediolateral columns

Anterior Spinal Artery Infarction



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Anterior Spinal Artery Infarction — Treatment

- Supportive

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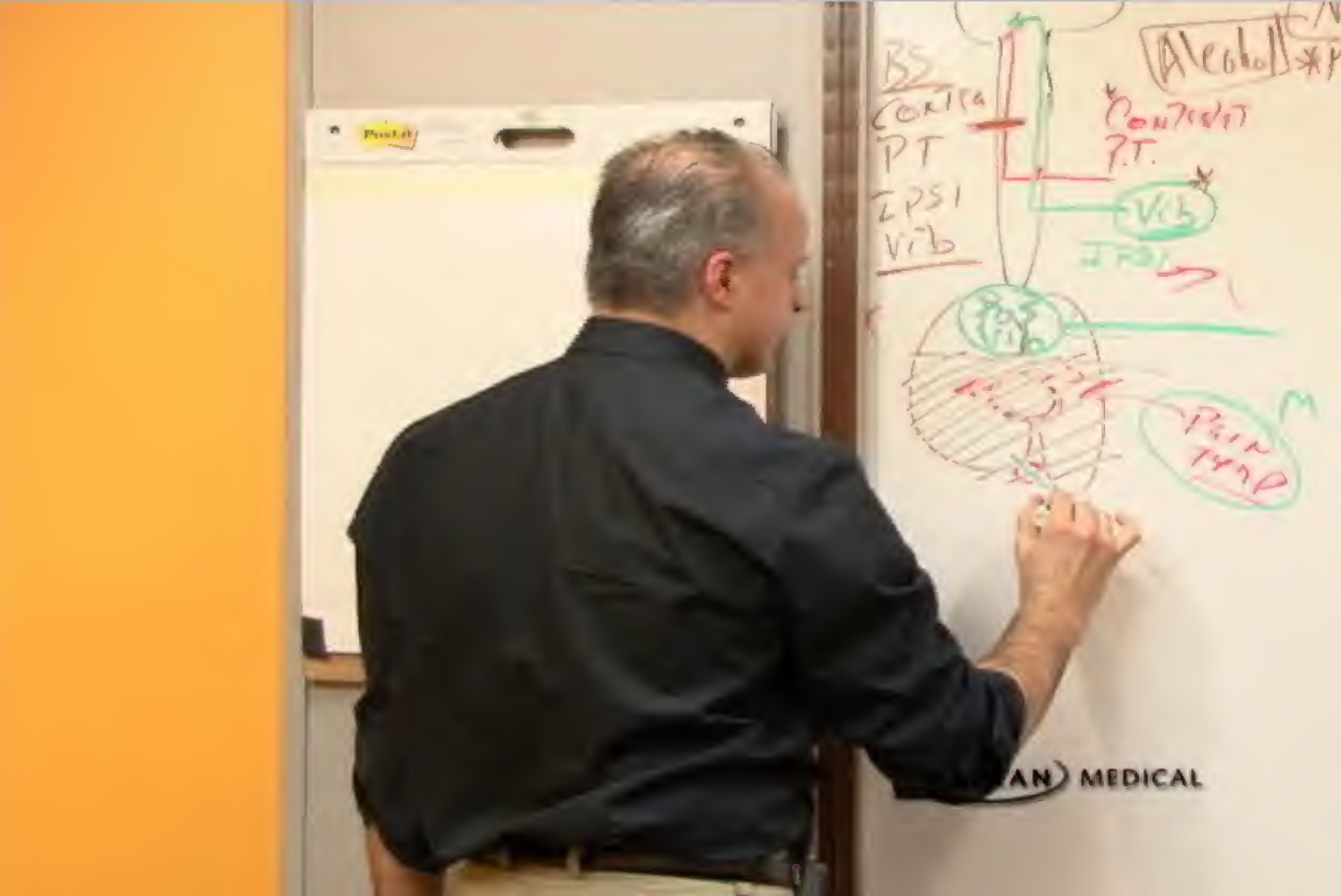
BS
CORTEX
PT
EPSI
Vib
Alcohol
Control
P.T.
Vib
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Anterior Spinal Artery Infarction

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Brown-Sequard Syndrome


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Brown-Sequard Syndrome

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Vertigo and Dizziness

Vertigo vs. Dizziness

Sensation	Diagnosis	Etiology
The environment is "spinning" Sensation of movement without actually moving	Vertigo	Neurological
Lightheadedness Feeling like they are going to "black out"	Presyncope	Cardiac

Central vs. Peripheral Disease

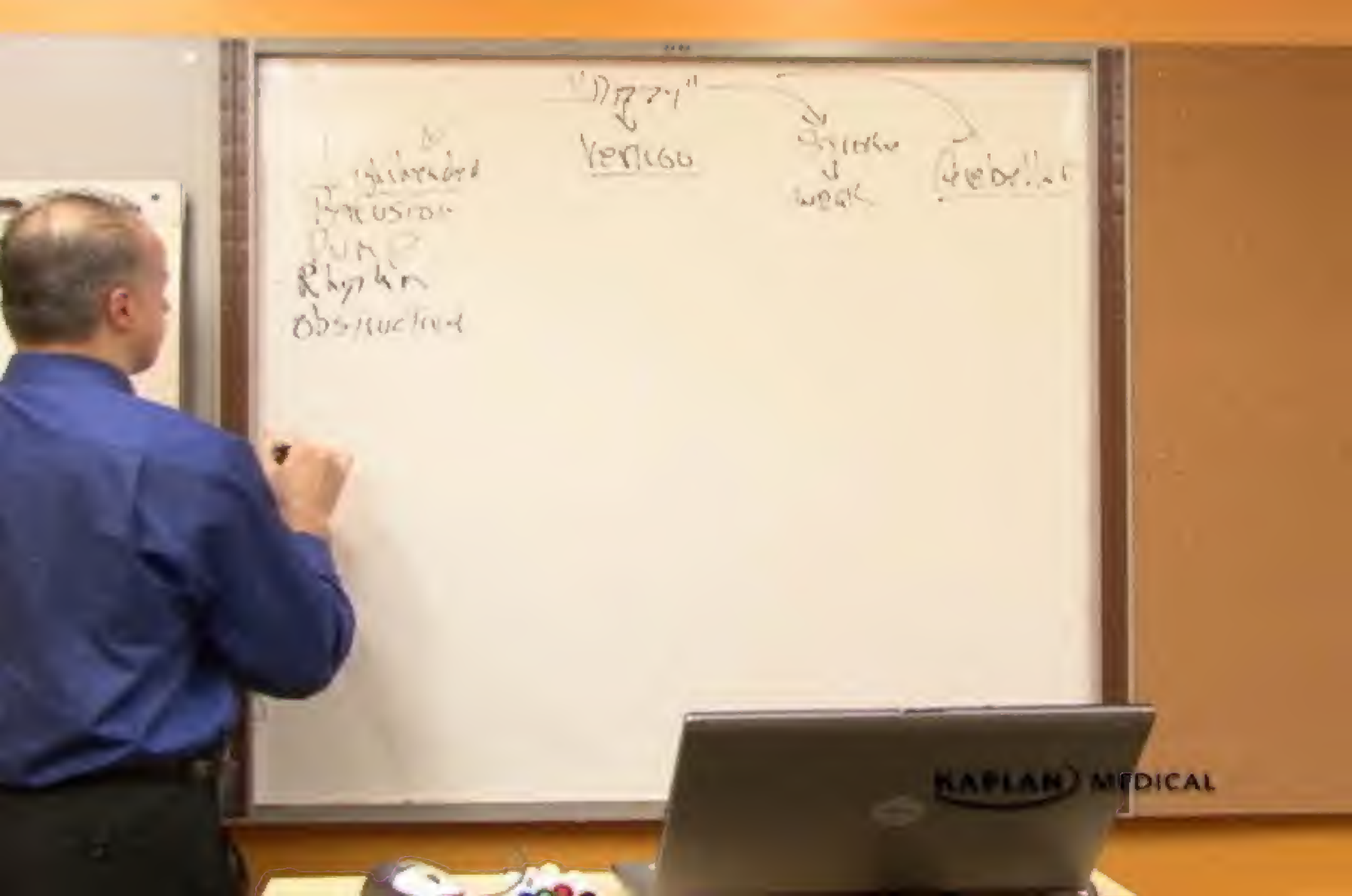
	Central Vertigo	Peripheral Vertigo
Onset	Gradual	Usually sudden
Tinnitus, Hearing Loss	Absent	Present
Neighborhood Signs (diplopia, cortical blindness, dysarthria, extremity weakness/numbness)	Present	Absent
Nystagmus	Pure, vertical, does not suppress with fixation, and multidirectional	Mixed, horizontal, suppresses with fixation, and unidirectional

Vertigo — Etiology

Peripheral Vertigo	• Meniere's Disease– tinnitus, hearing loss and episodic vertigo lasting 1-8 hours • Labyrinthitis– sudden and severe, lasts days with hearing loss and tinnitus • Benign paroxysmal positional vertigo– exacerbated by movement lasting seconds • Perilymphatic fistula– due to trauma

Vertigo — Treatment

- Based on etiology
- Peripheral vertigo and labrynthitis
  - Symptomatic treatment with meclizine or diazepam when severe
- Meniere's Disease
  - Low-salt diet and diuretics, surgery if failure to medical therapy occurs
- Benign paroxysmal positional vertigo
  - Positional maneuvers to dislodge the otolith


Vertigo
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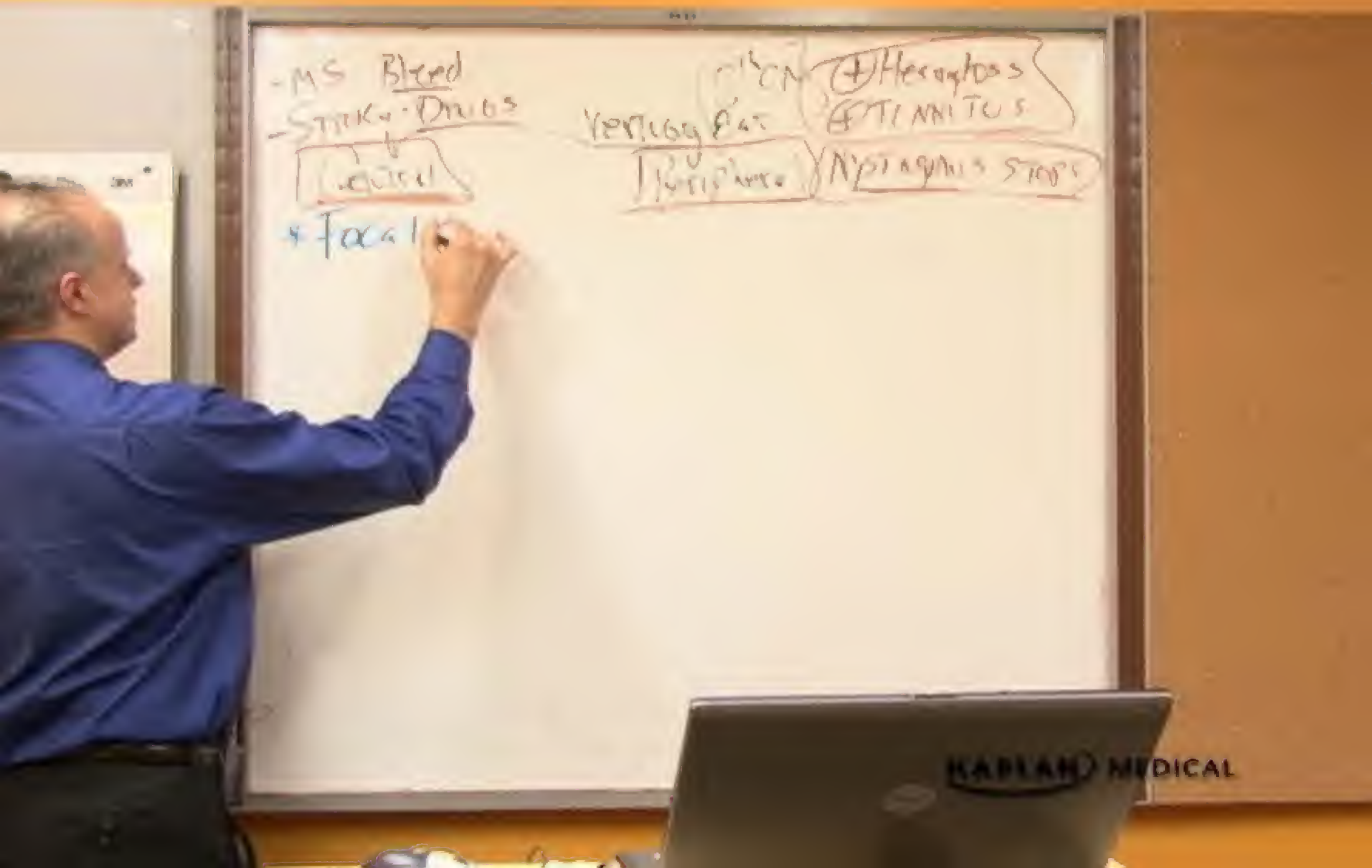
-MS Bleed
-Stroke - Drugs
Vertigo
Hearing
Nystagmus stops
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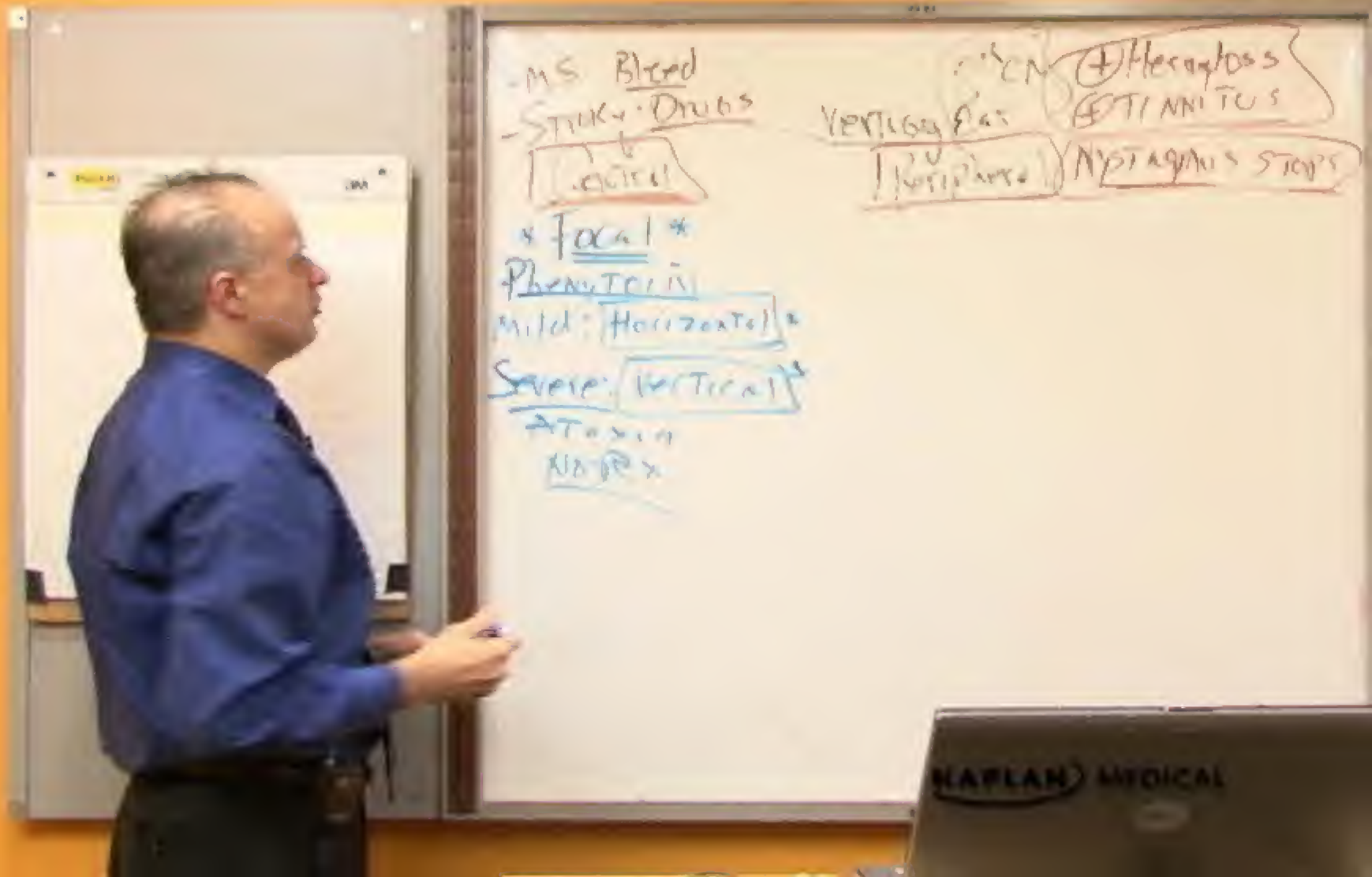
- MS Blood
- Sticky Drugs
Focal
Phenytoin
Mild: Horizontal
Severe: Vertical
Toxin
Vertigo/Gas
Hemoptosis
TINNITUS
Nystagmus stops
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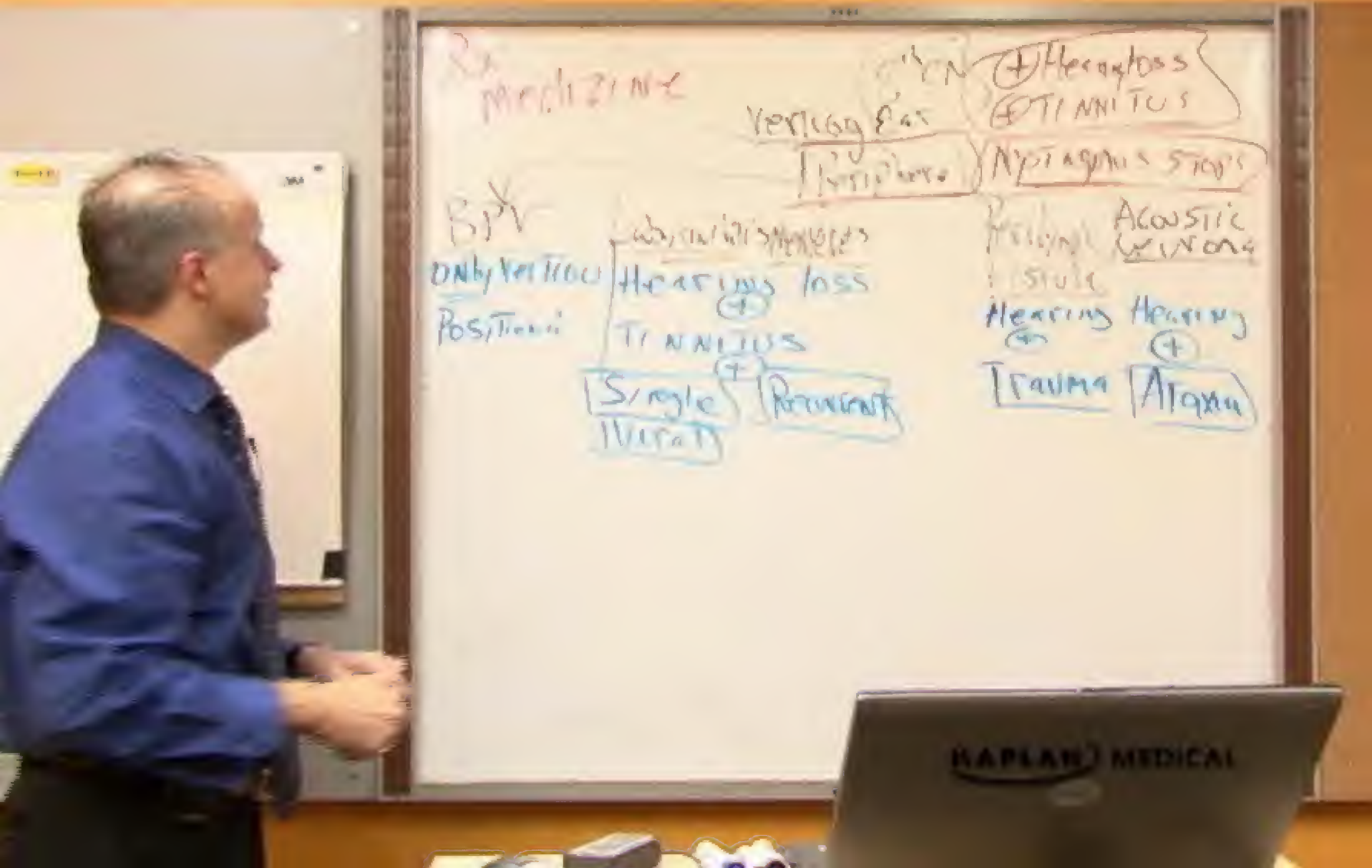
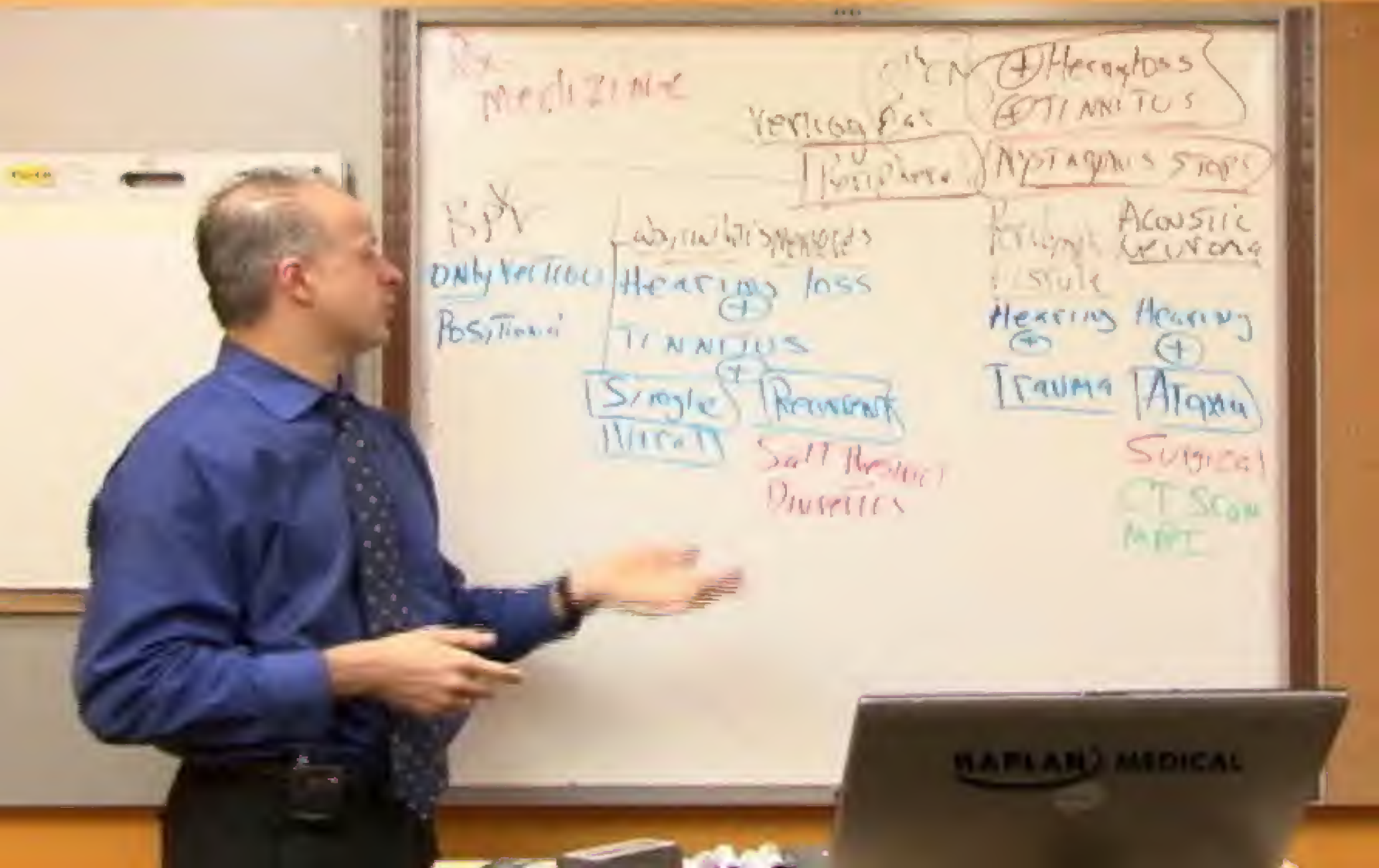

Vertigo Ear
Hearing loss
TINNITUS
Hearing loss
TINNITUS
Single
Recurrent
Trauma
Atoxic
KAPLAN MEDICAL


MEDICINE
Vertigo
Hearing loss
TINNITUS
Single
Recurrent
Hearing
Trauma
Ataxia
Surgical
CT Scan
MRI
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Vertigo and Dizziness

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Headache

Headache — Etiology

Primary Headaches	***Secondary Headaches***
Migraines	Intracranial hemorrhage
Cluster headache	Brain tumor
Tension headache	Meningitis
	Temporal Arteritis
	Glaucoma

Headache — Differential Diagnosis

- Primary Headaches
  - Usually recurrent
- Secondary Headaches
  - Usually sudden or severe and accompanied by worrisome signs and symptoms

Headache — Differential Diagnosis

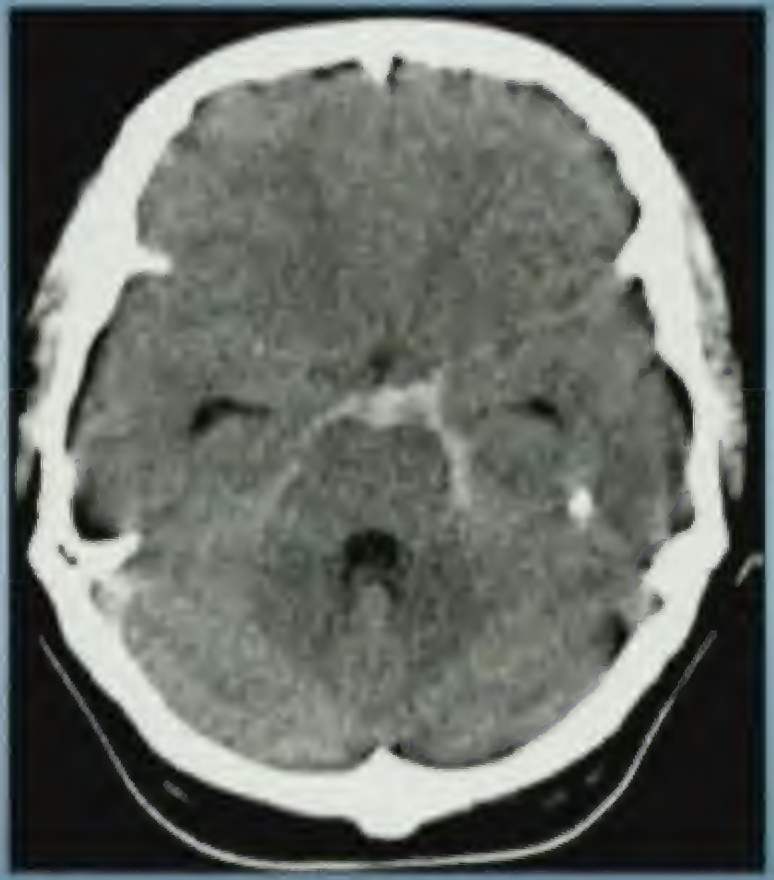
Meningitis	Fever and nuchal rigidity
Intracranial hemorrhage	"Worst headache of my life"
Brain tumor	Deep, dull, aching, wakes them from sleep
Posterior fossa tumors	Vomiting precedes headache by weeks. Worse with bending, lifting or coughing
Temporal arteritis	Unilateral pounding headache with lancinating pain and visual changes

Headache — Differential Diagnosis

- Once serious pathology (secondary causes) are ruled out, proceed to investigate primary causes

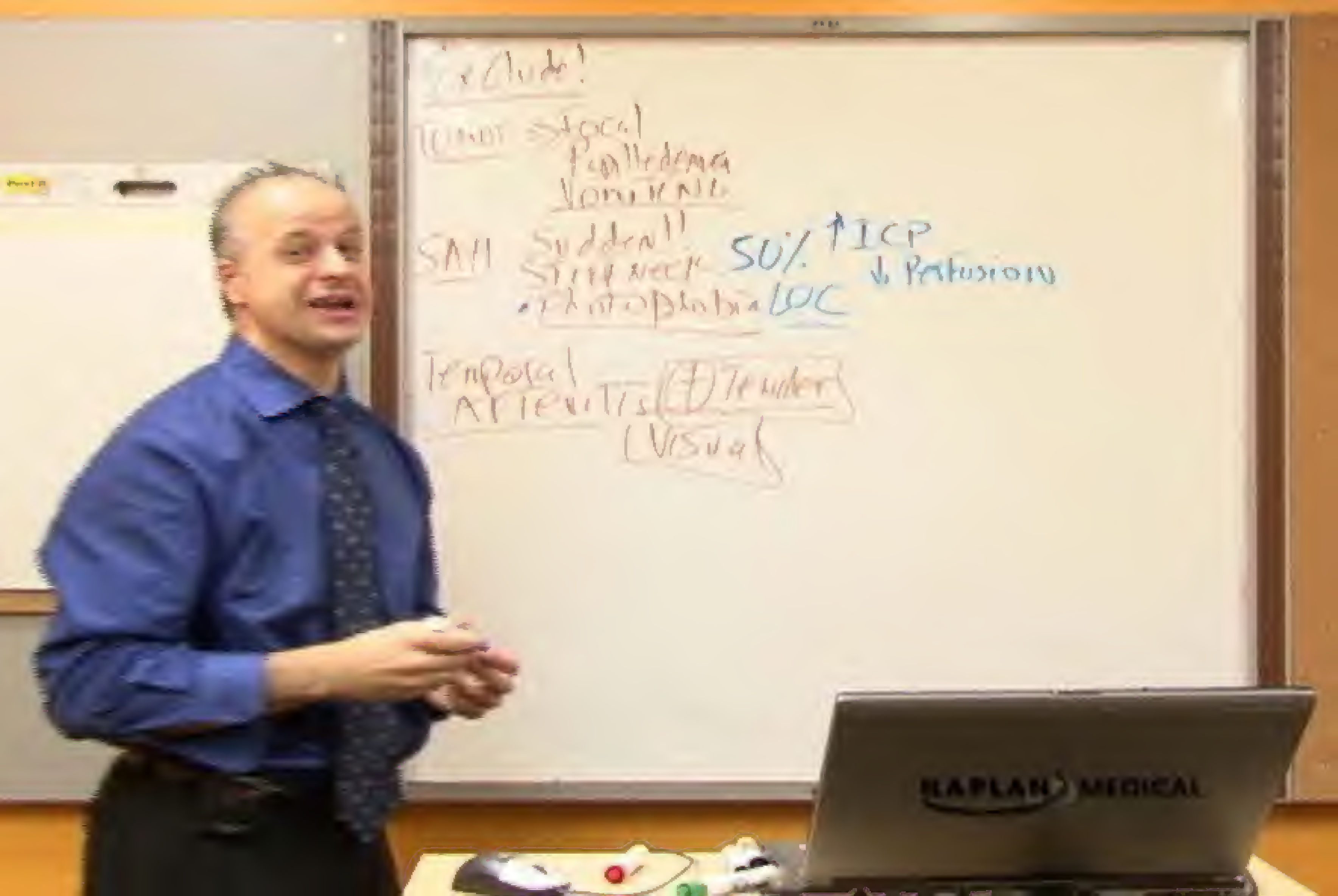
Migraine headache	Pulsatile, throbbing, unilateral, aggravated by movement, light or sound
Tension headache	Tight, band like, bilateral with tightness of the posterior neck muscles; builds slowly and may last days
Cluster headache	Excruciating, unilateral, periorbital lasting up to 90 mins. Associated with rhinorrhea, red eye, lacrimation, nasal congestion and nausea

“The worst headache of my life”



Migraine Headache — Treatment

Abortive Treatments	***Prophylactic Treatments***
NSAIDs, aspirin, acetaminophen Triptans Ergotamine derivatives	Beta-blockers Calcium channel blockers Tricyclics SSRIs Valproic acid Topiramate


Papilledema
Vomiting
SAH
Sudden!!
Stiff neck
Photophobia
50%
↑ICP
↓ Perfusion
LOC
Temporal Arteritis
(+) Tender
Visual
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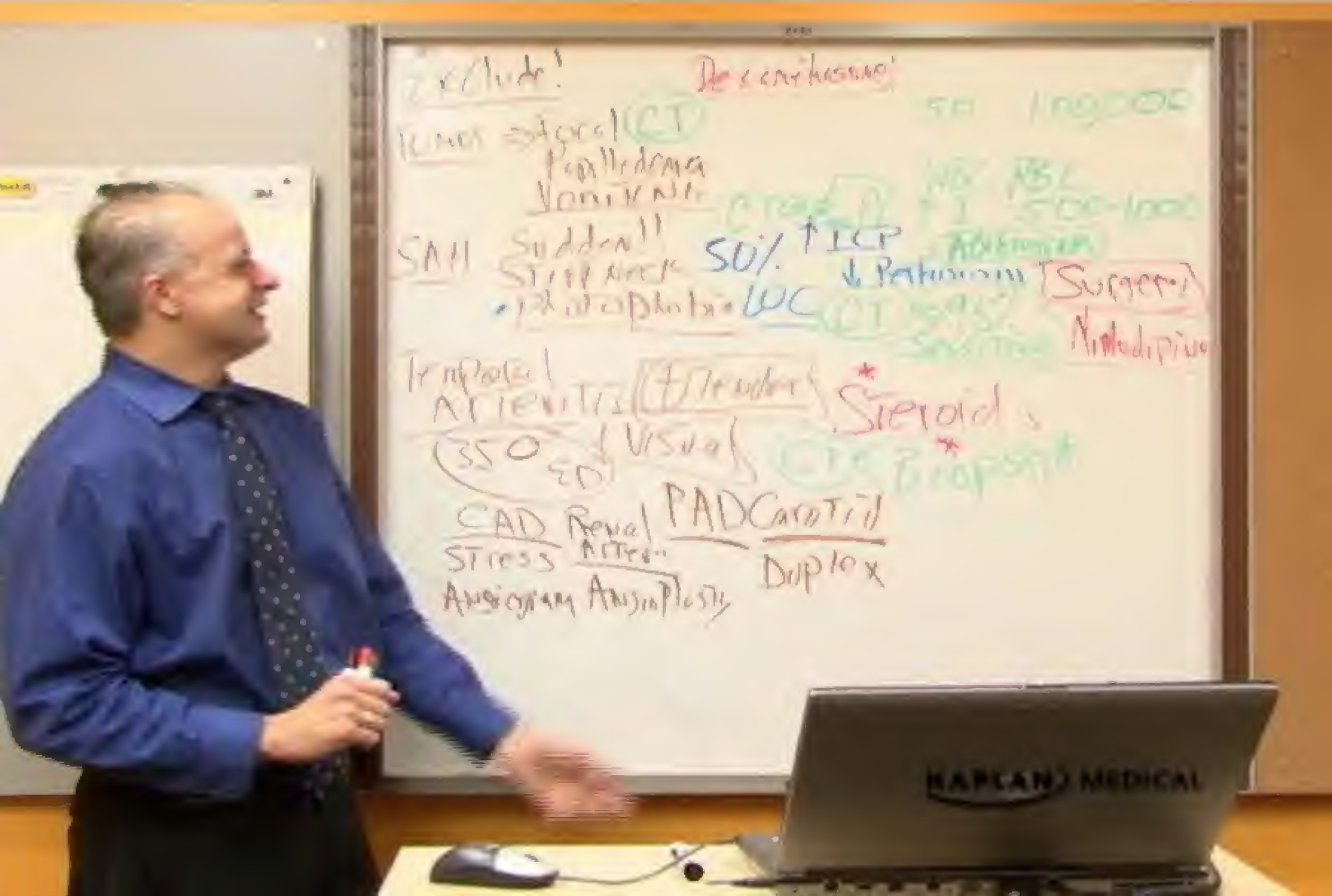
Sterioid
PAD Carotid
Duplex
CAD
Stress
Angiogram
Angioplasty
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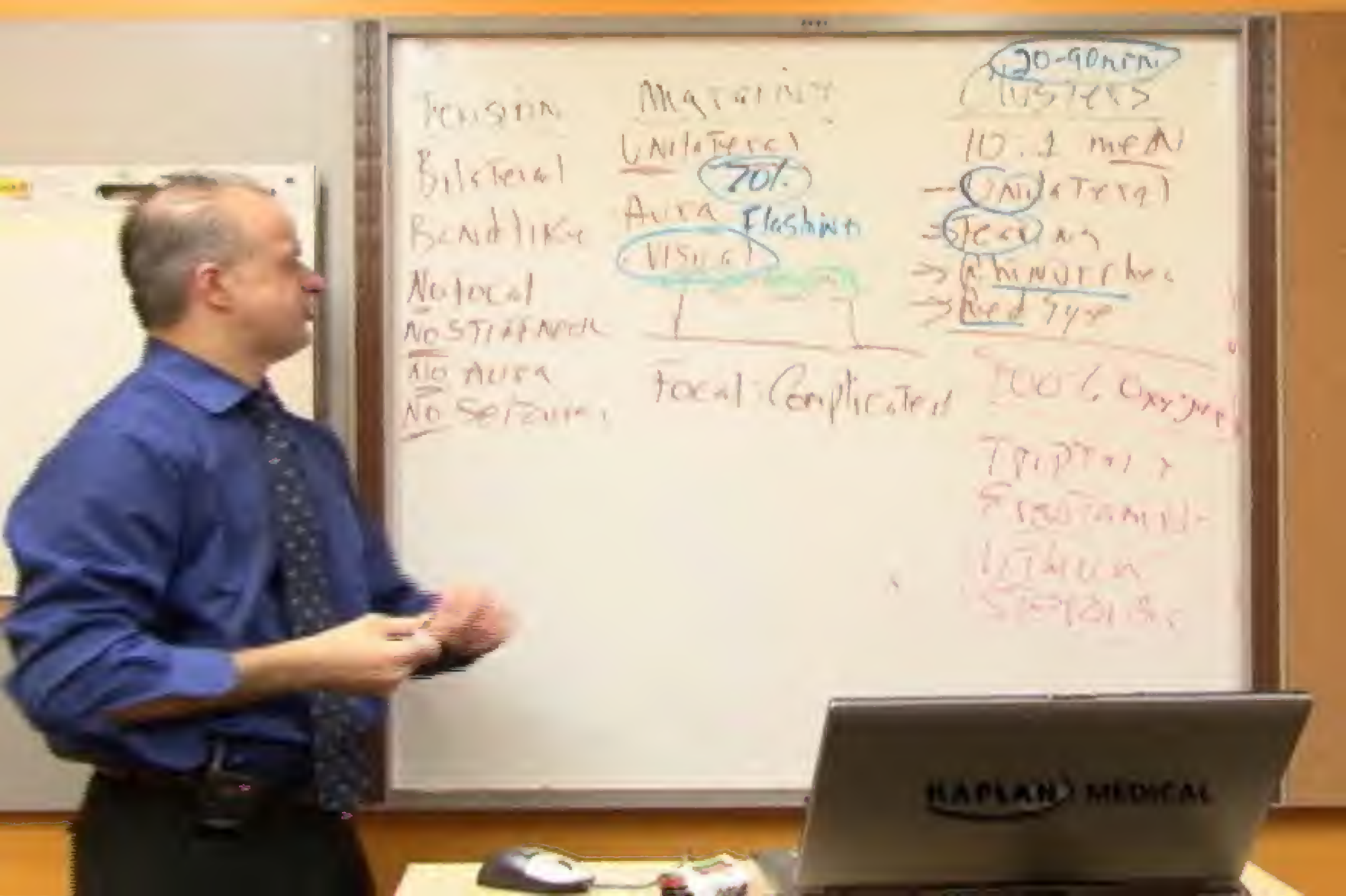
Tension
Bilateral
Bandlike
Notocal
NO STIFFNESS
No Aura
No Seizures
Migraine
Unilateral
70%
Aura Flashing
Visual
Focal Complicated
20-90min
Clusters
10:1 men
Unilateral
Tears
Rhinorrhea
Red Eye
100% Oxygen
Triptan
Prednisone
Lithium
Steroids
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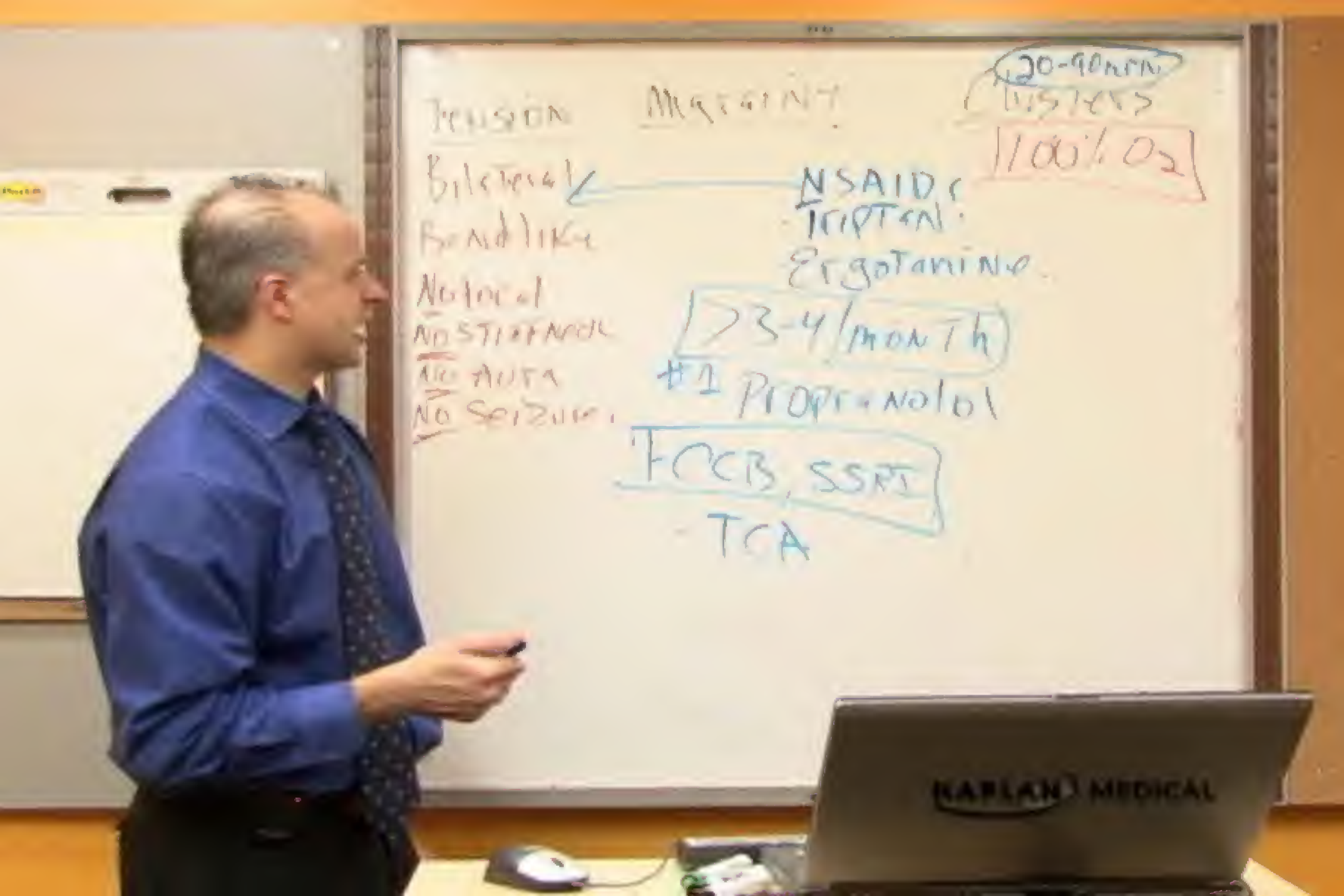
TENSION
MIGRAINE
20-90 MIN
CLUSTERS
100% O2
Bilateral
Band like
Notical
NO STEP AND
NO AURA
NO SEIZURE
NSAIDS
TRIPTANS
Ergotamine
>3-4/MONTH
#1 PROPRANOLOL
FCCB, SSRI
TCA
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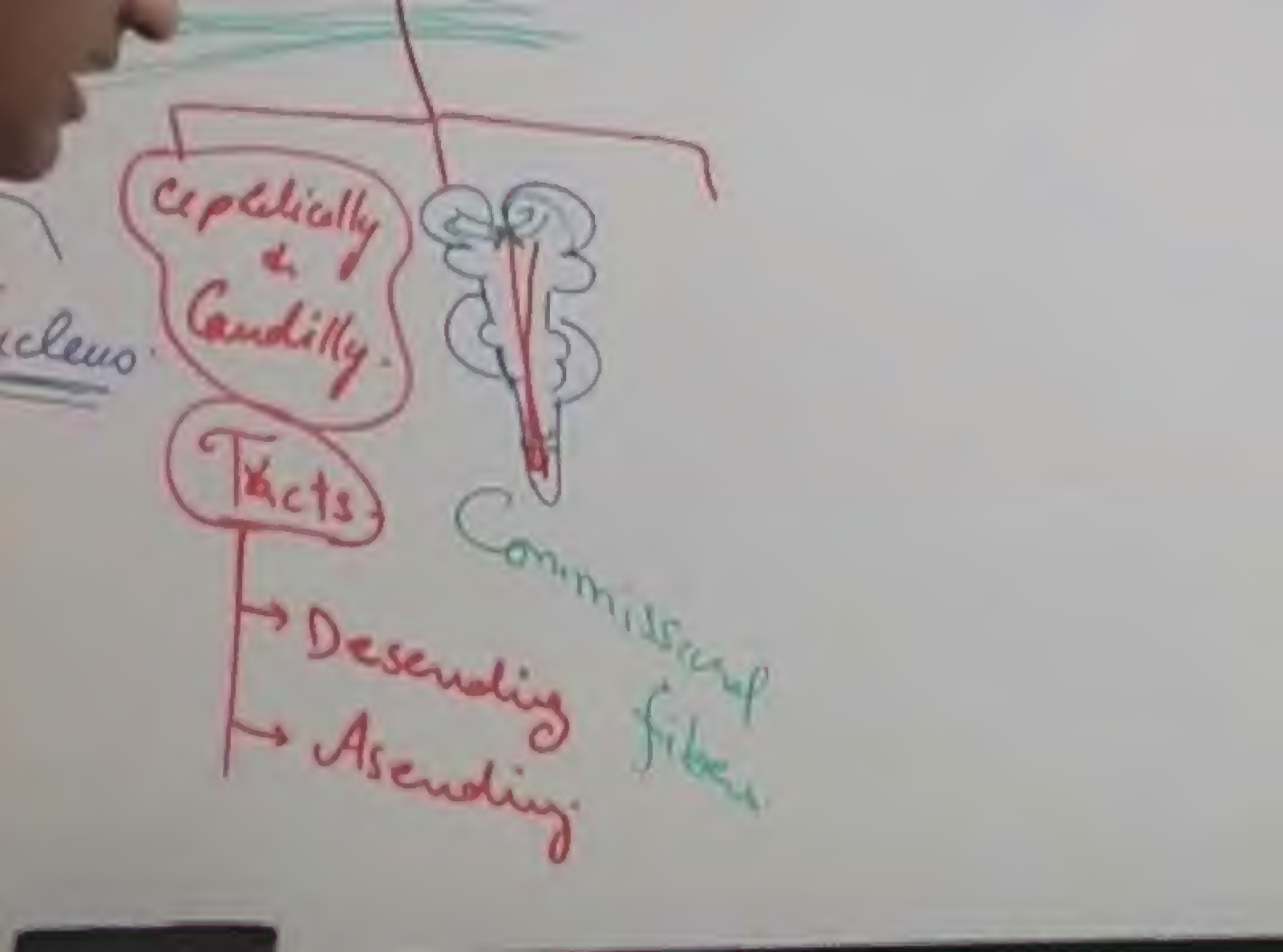
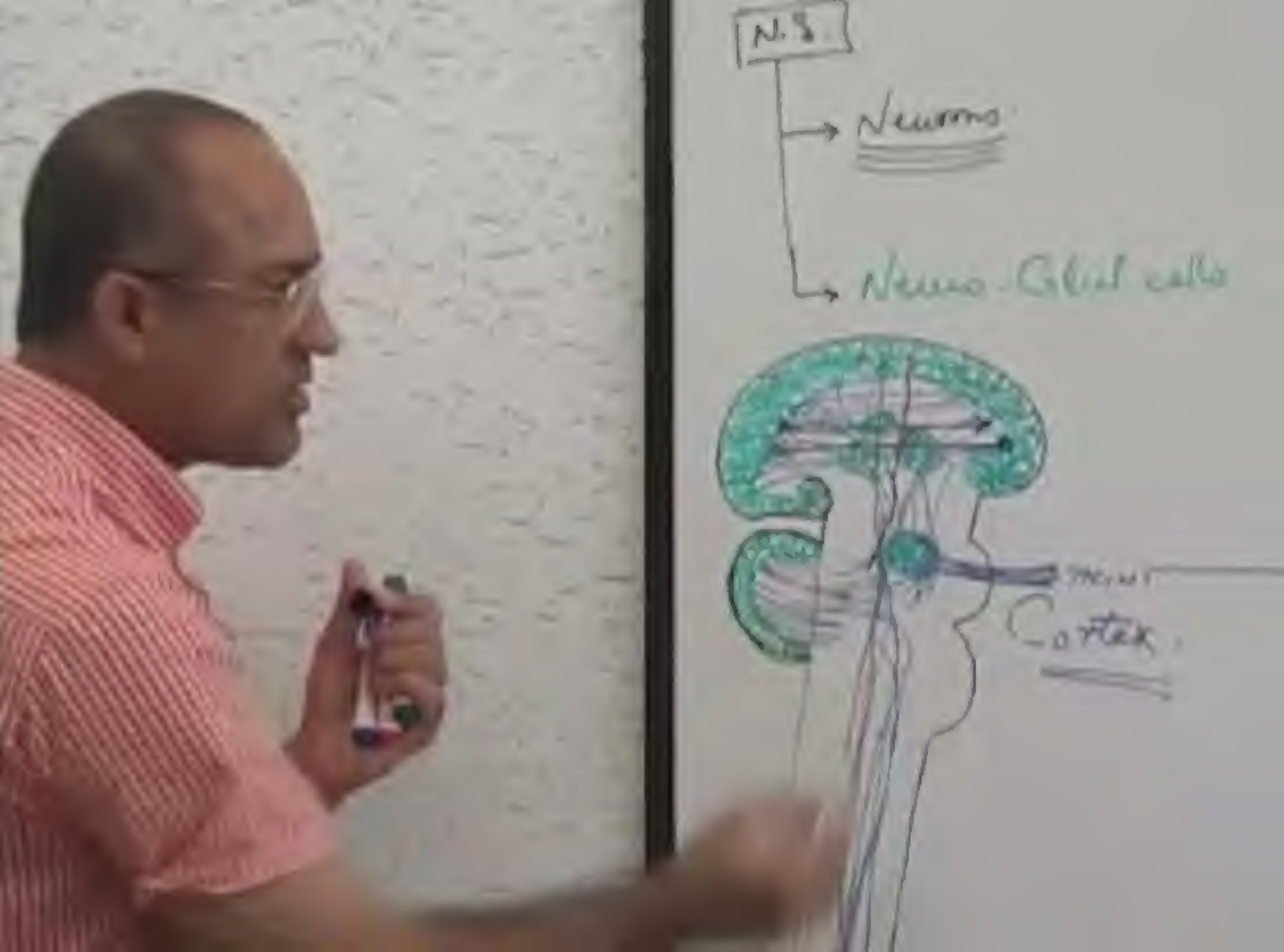
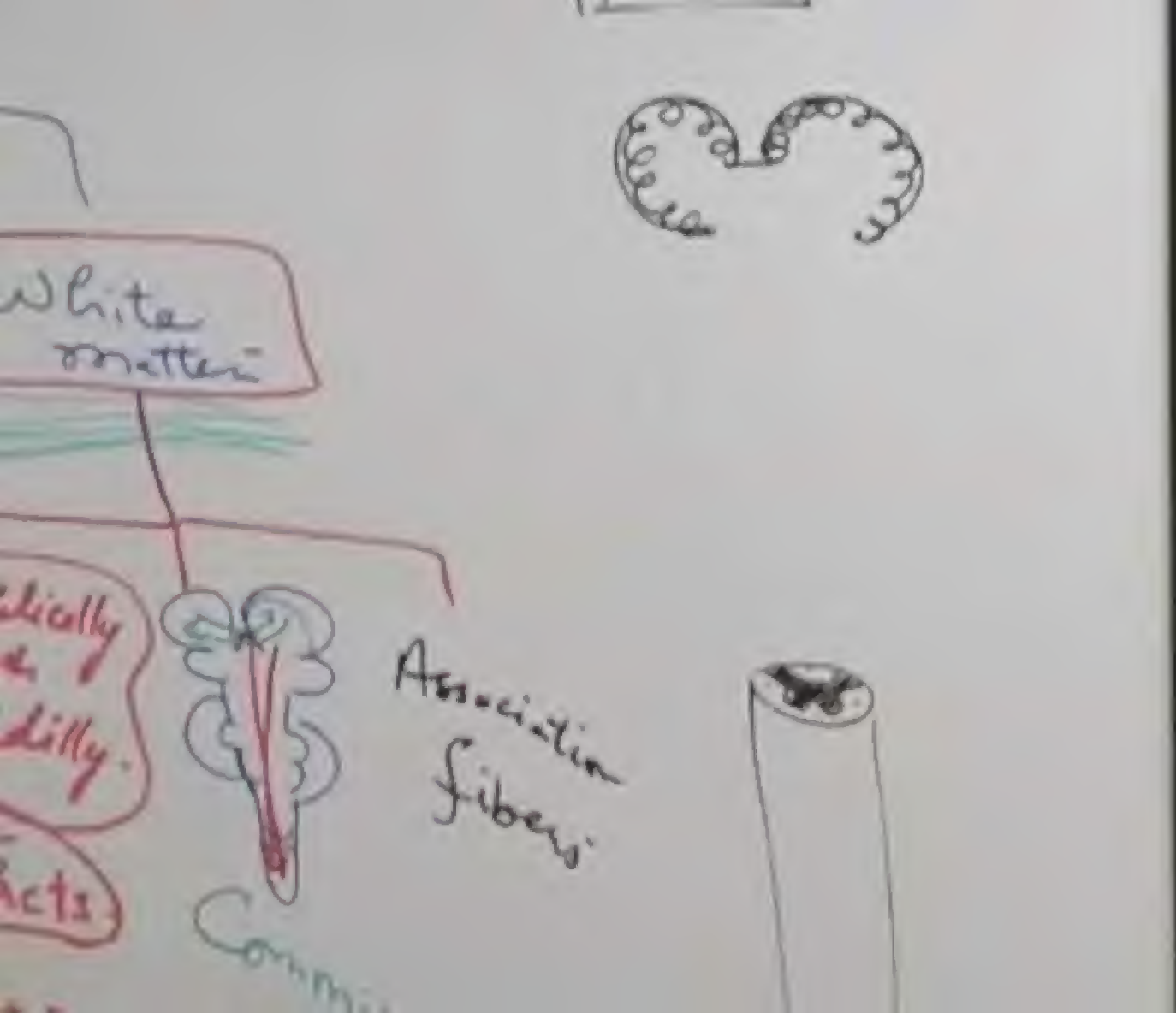
Headache

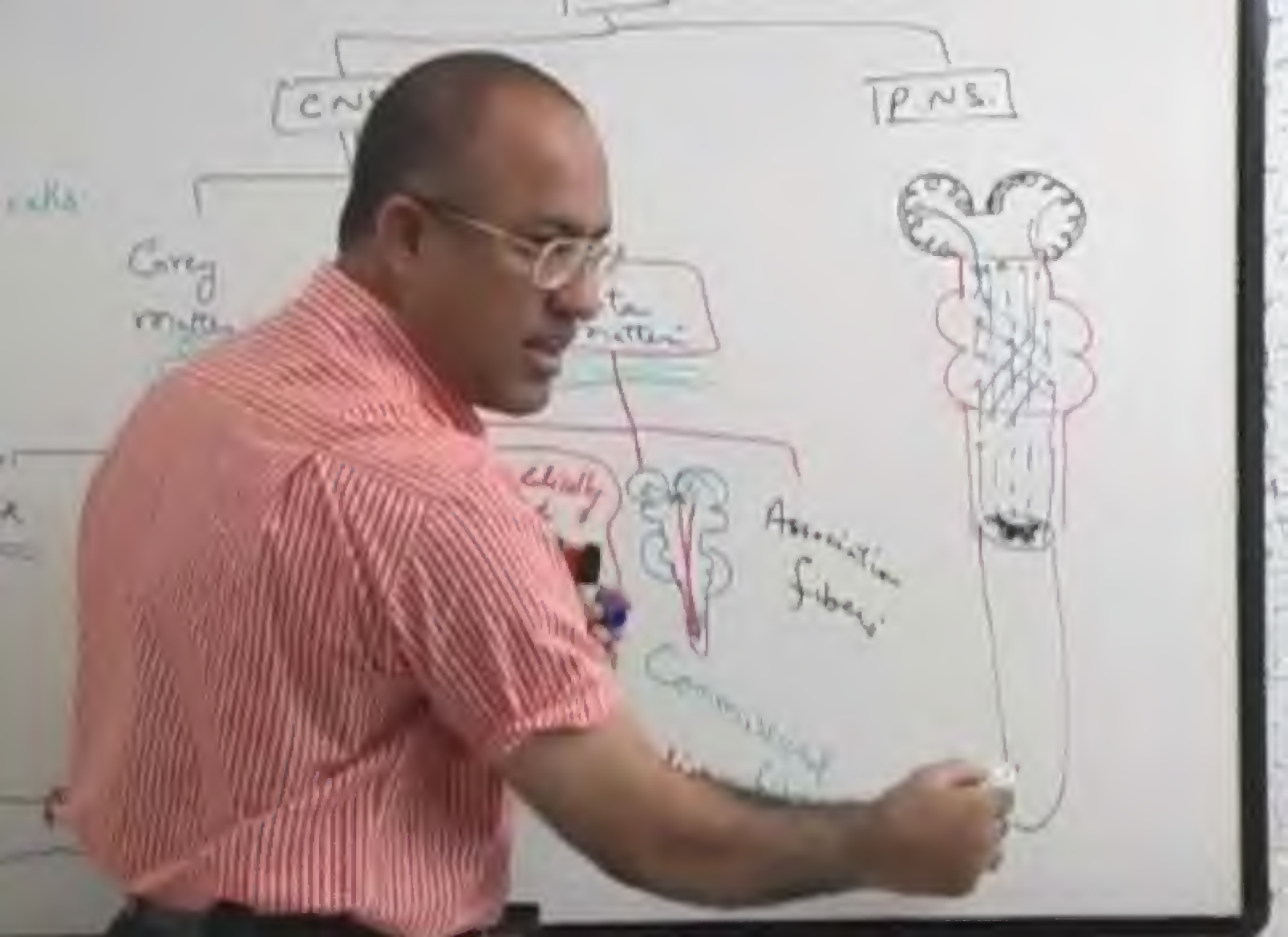
END

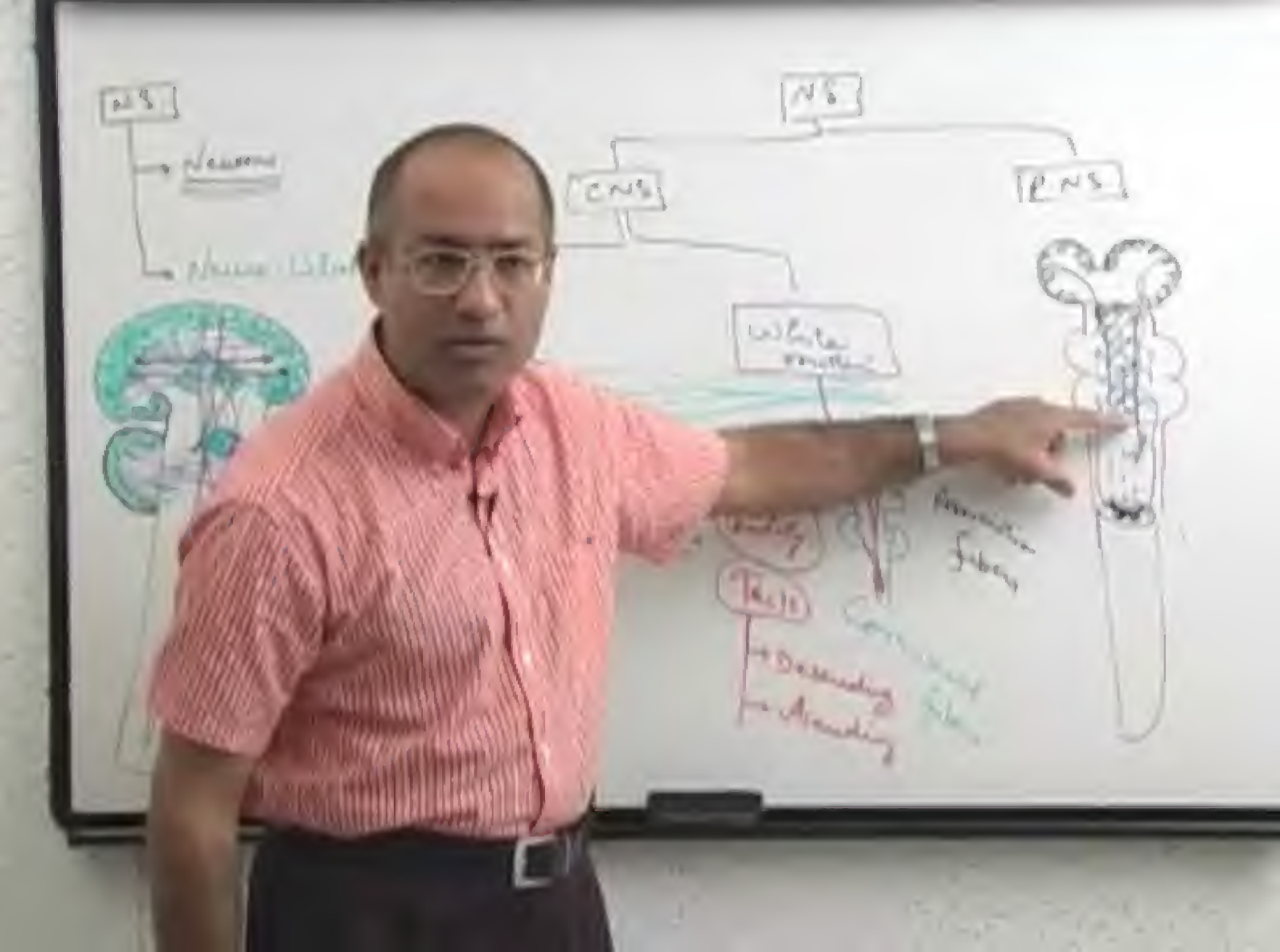

C.N.S
P N S
Sensory PNS
Special Sensation
General Sensation

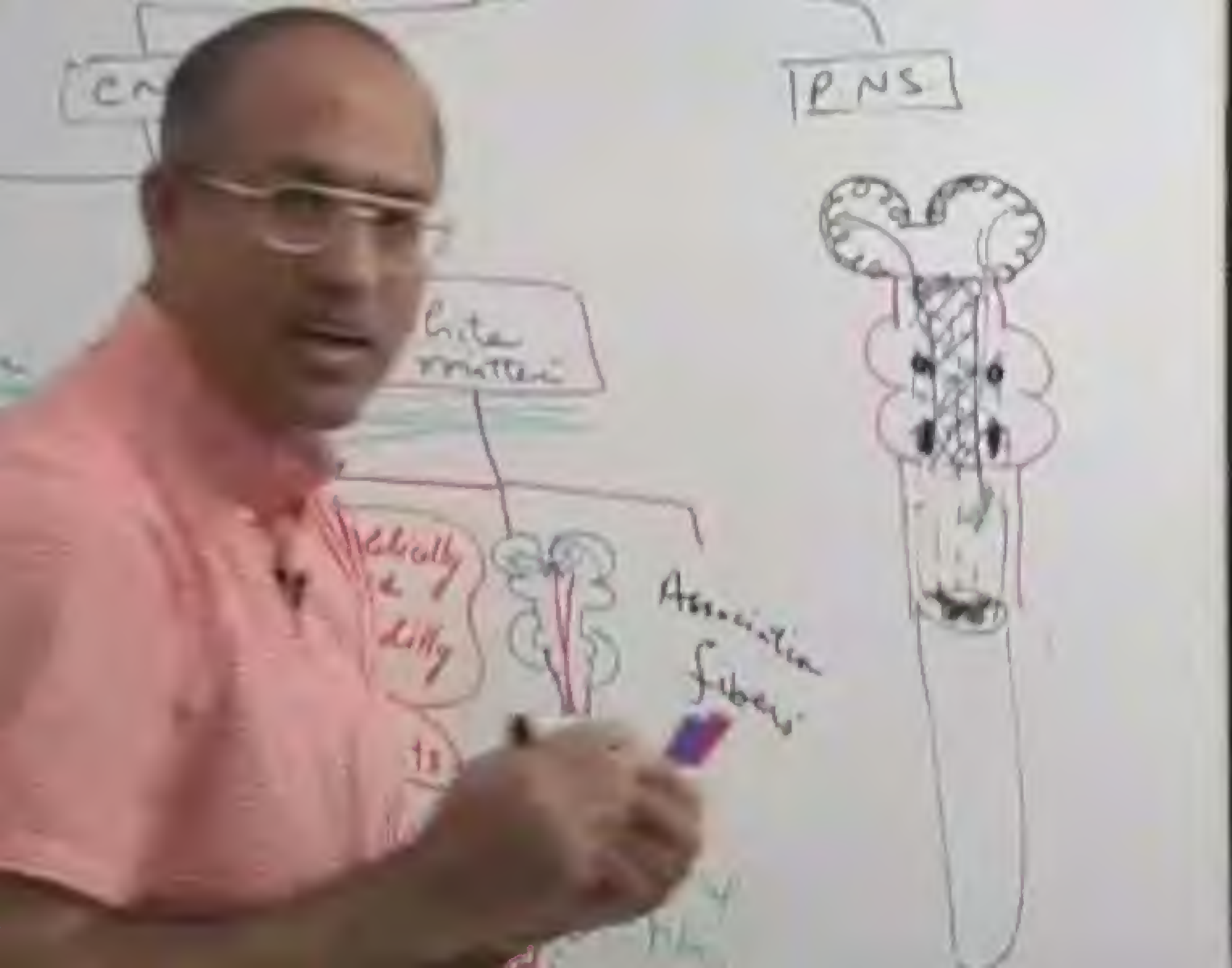

Superficially & Candilly.
Tracts
Descending
Ascending
Commissural fibers


N.S.
Neurons
Neuro Glial cells
Cortex


White matter
Association fibers


P.N.S.
Association fibers


N S
CNS
PNS


PNS
Association
fibers

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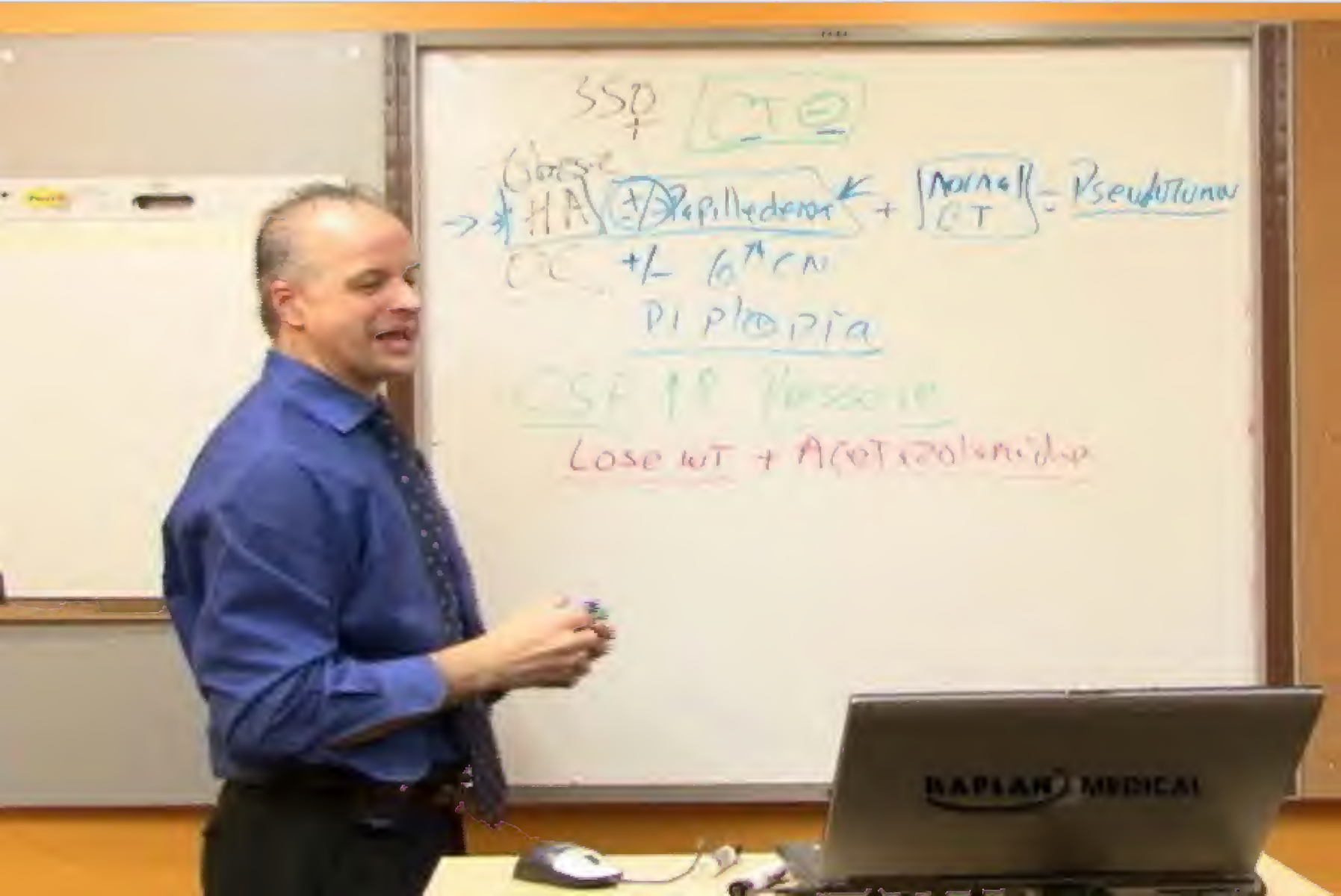
Pseudotumor Cerebri


SSD
CT
HA
Repilledores
Normal
CT
Pseudotumor
+/- 6ª PN
DIPLOPIA
Lose WT + Acetazolamida
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Pseudotumor Cerebri

End

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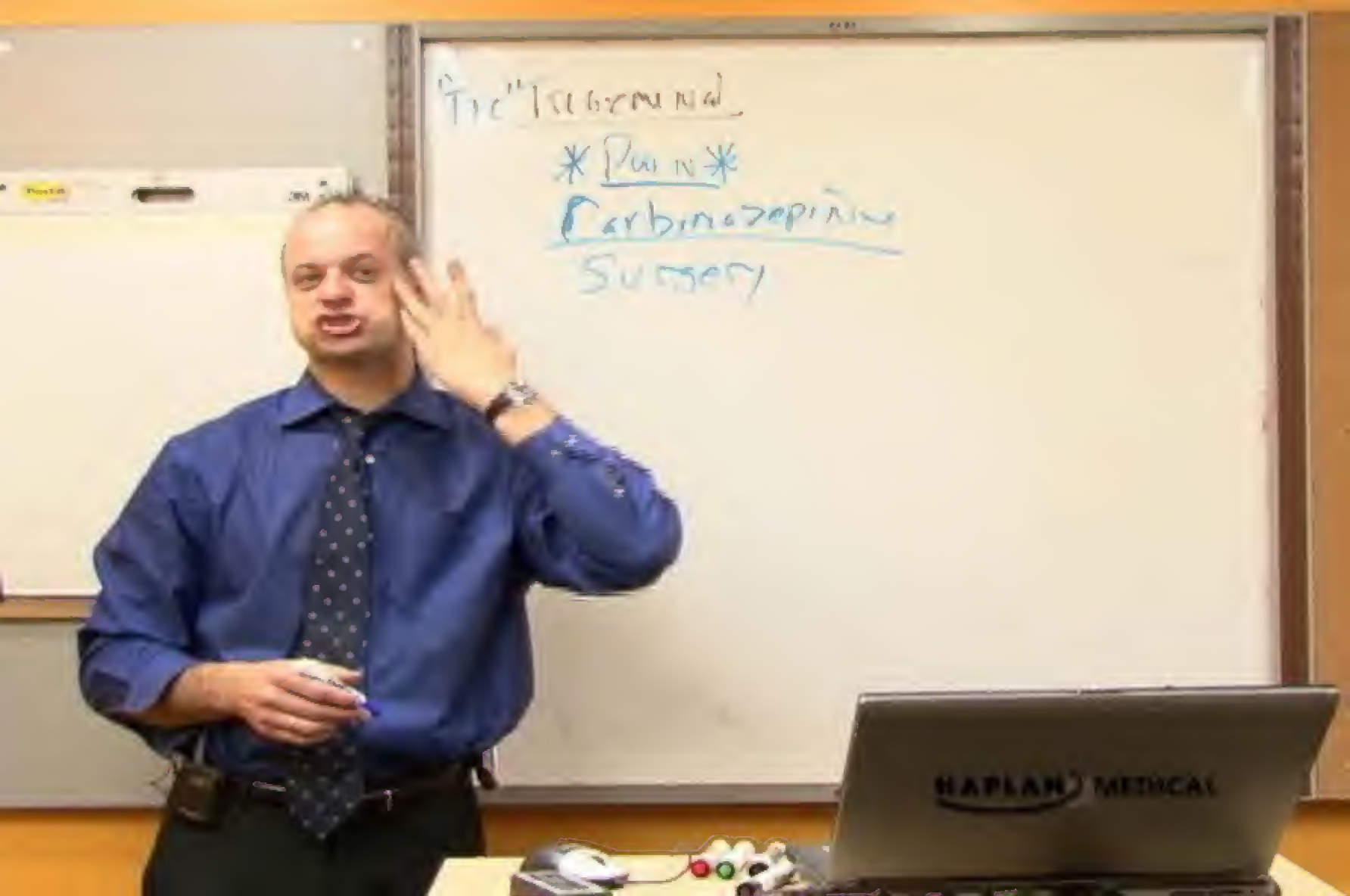
Trigeminal Neuralgia


Tic Treatment
* Pain *
Carbamazepine
Surgery
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Trigeminal Neuralgia

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Ch. 5: Guillian Barrè Syndrome

Guillian Barrè Syndrome — Etiology

- Autoimmune destruction of myelin
- History of infection 1-3 weeks prior to onset
  - Respiratory or GI system
  - *Campylobacter jejuni*, human herpesvirus, cytomegalovirus and Epstein-Barr virus have all been implicated
- Due to molecular mimicry: the immune system attacks self-antigens as foreign-antigens

Guillian Barrè Syndrome — Presentation

- Rapid development of weakness starting in the lower extremities and moving upwards
- Absent reflexes
- Progression over hours to days
- Legs > arms
- Pain and tingling sensations
- Presence of fever, constitutional symptoms or bladder dysfunction should raise questions to the diagnosis

Guillian Barrè Syndrome — Diagnosis

- Best initial test ⟹
  Lumbar puncture
  - Changes occur >48 hours after onset
  - Increased protein without increased cell count
- Most accurate test ⟹
  Electromyelography (EMG)
  - Detects demyelination of peripheral nerves

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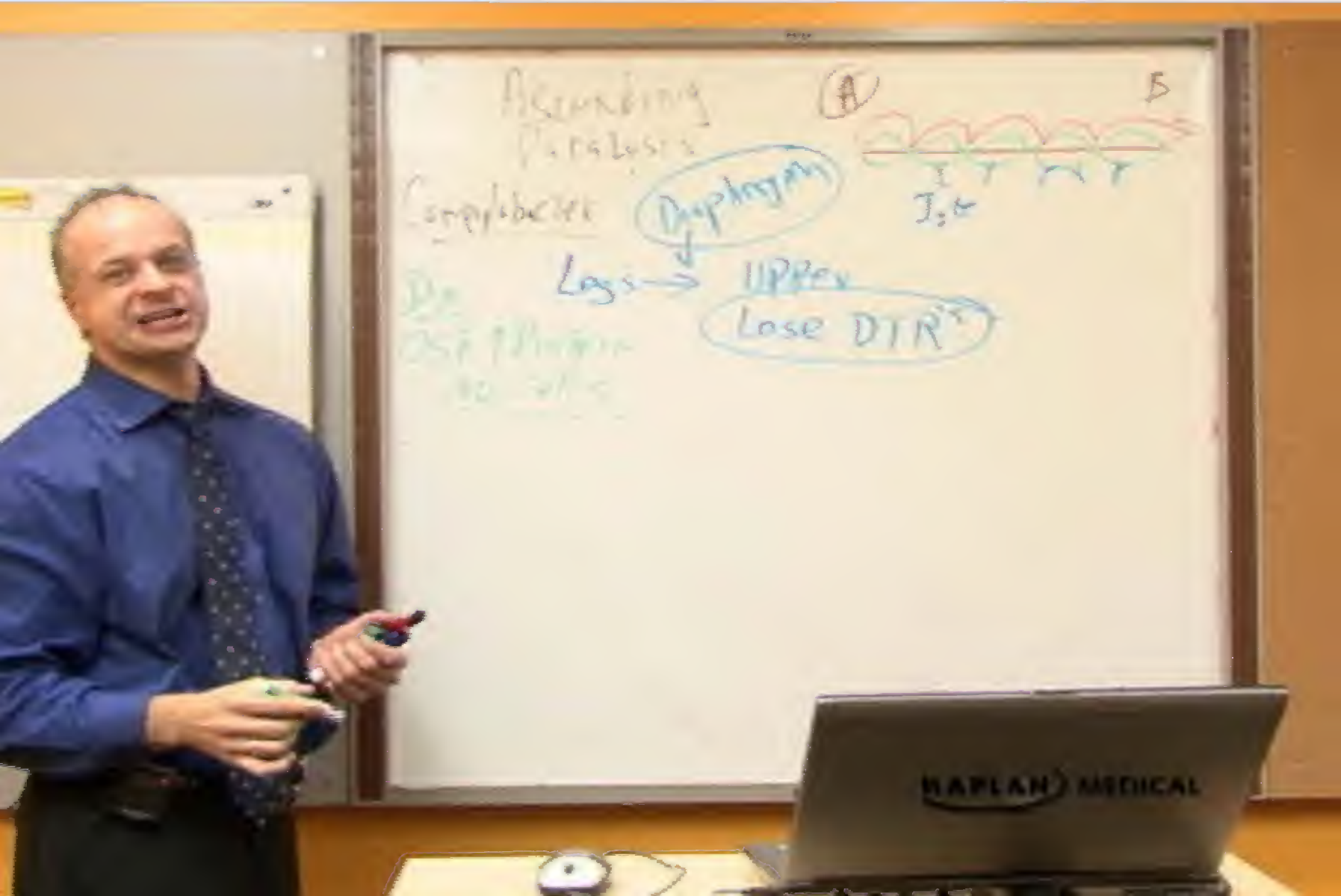
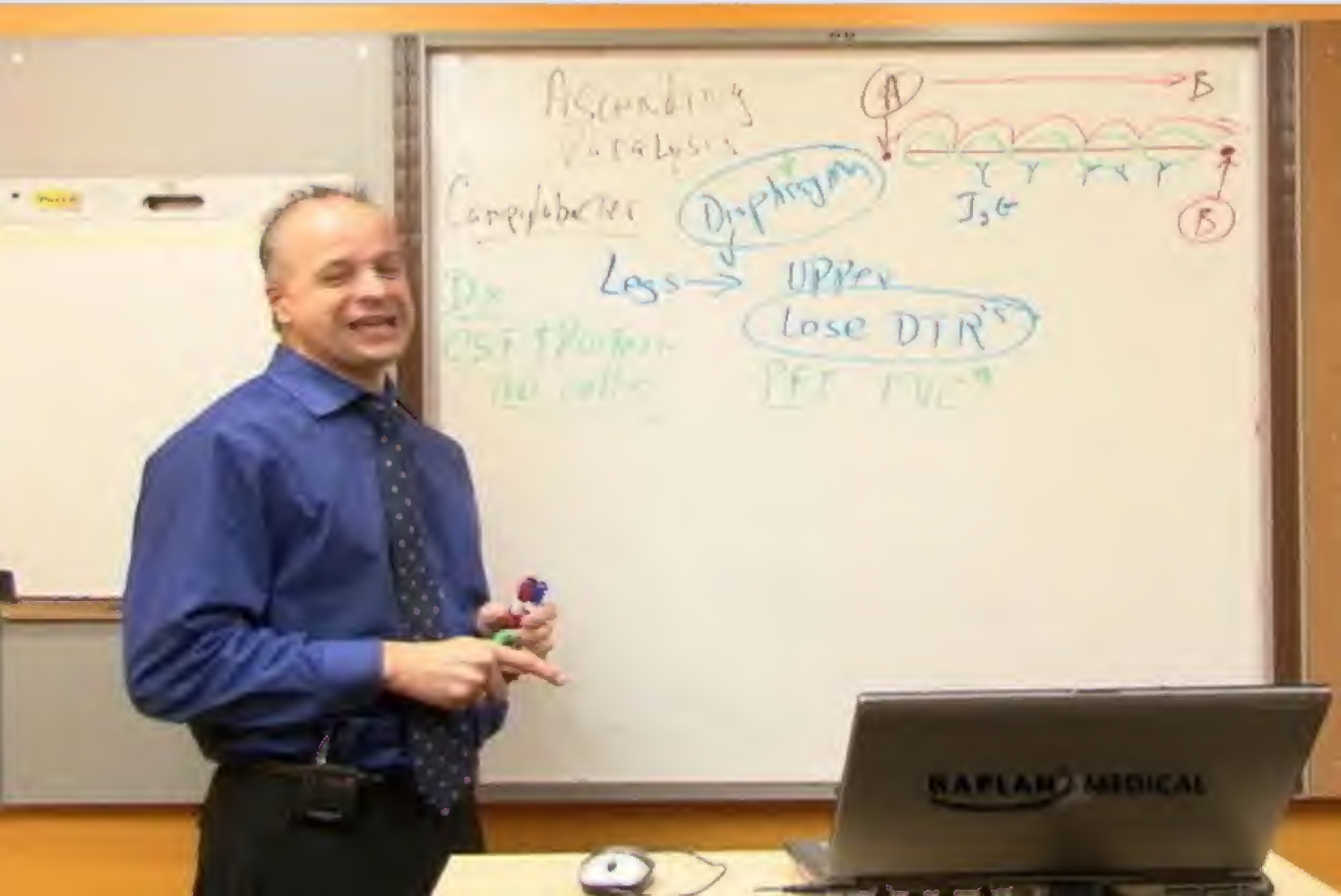
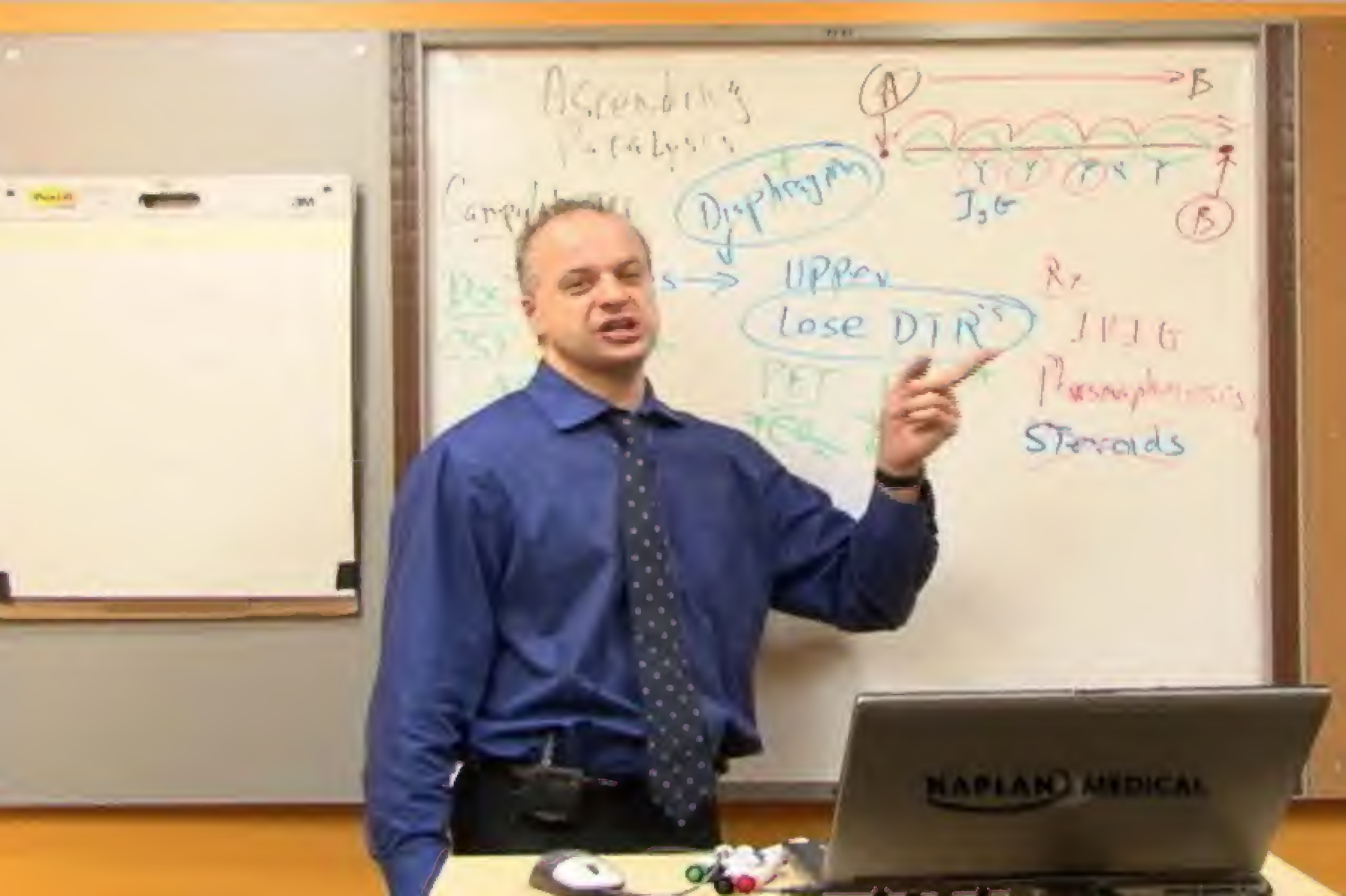
Guillian Barrè Syndrome — Treatment

- Should be started ASAP
  - Becomes ineffective 2 weeks after onset of symptoms
- Intravenous immunoglobulin or plasmapheresis
  - Both are equally effective
- Monitor for impending respiratory failure and intubate with mechanical ventilation if required


A
B
Diphtheria
Logs
Lose DTR
KAPLAN MEDICAL


A
B
Diaphragm
Loss
Lose DTR
RAPLAN MEDICAL


A
B
Diaphragm
lose DTR's
STeroids
NAPLAN MEDICAL

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Ch. 5: Guillian Barrè Syndrome

END

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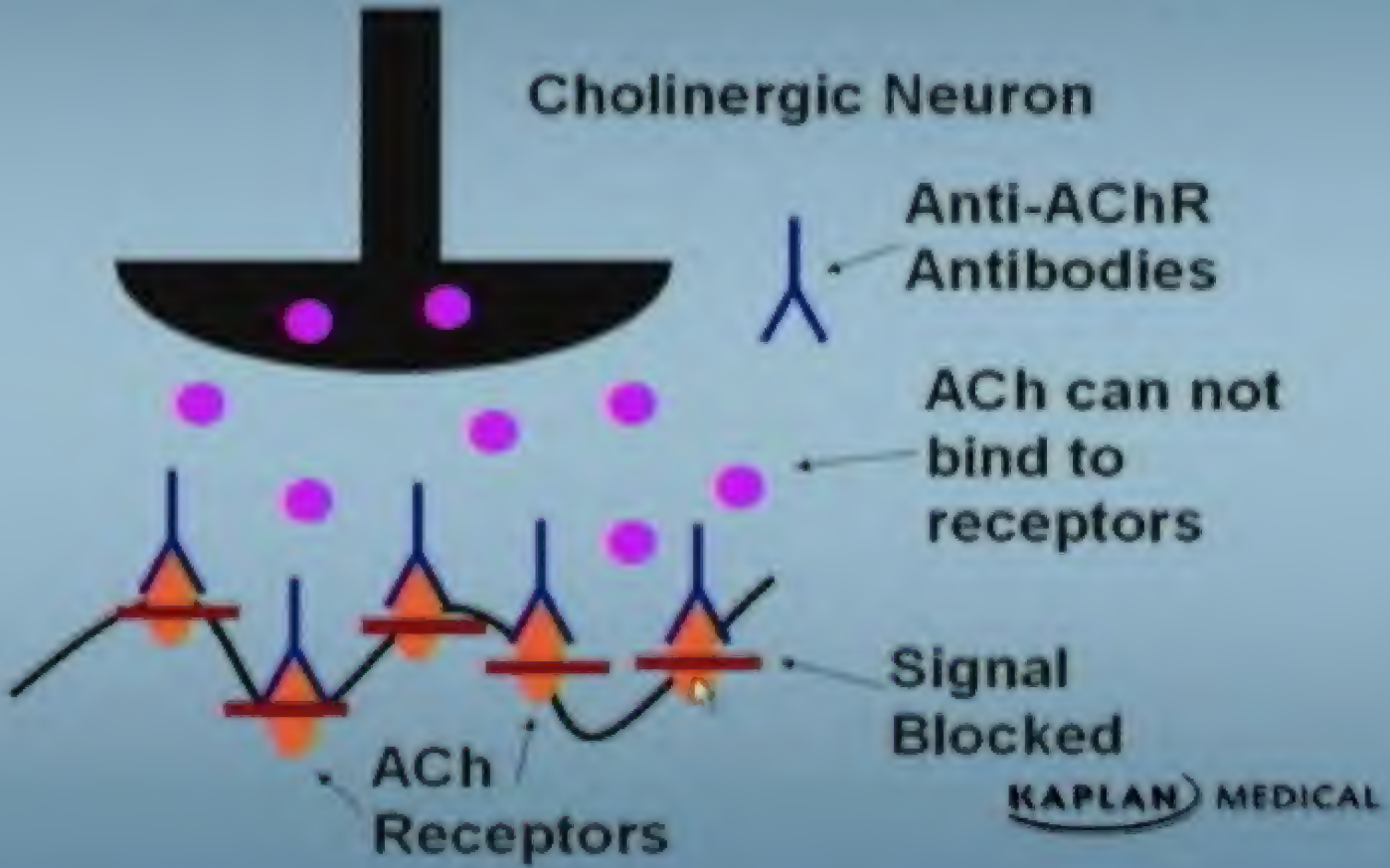
Ch. 6: Myasthenia Gravis

Myasthenia Gravis — Etiology

Acetylcholine auto-antibodies directed at the neuromuscular junction

Decreased number of active and functional acetylcholine receptors at the post-synaptic membrane

Myasthenia Gravis — Etiology



Myasthenia Gravis — Clinical Presentation

- Diplopia, ptosis and difficulty swallowing
- Nasal speech
- “Snarling” smile
- Weakness may become generalized and asymmetric
- Deep tendon reflexes are intact, no sensory abnormalities, normal pupillary light reflex

Myasthenia Gravis — Differential Diagnosis

- Eaton-Lambert Syndrome
  - *Increasing* muscle strength with repetitive movement
  - Associated with small cell carcinoma of the lung

Myasthenia Gravis — Diagnosis

- Best initial test
  - Acetylcholine receptor antibody test
  - Virtually diagnostic with generalized symptoms (80-90%)
  - Less diagnostic with disease limited to the eyes (70%)
- Edrophonium (Tensilon®) test
  - Sensitive but not specific

Myasthenia Gravis — Diagnosis

- Chest X-Ray
  - Rule out thymoma
- Most Accurate test
  - EMG (electromyelography)- decremental decrease in muscle fiber contraction on repetitive nerve stimulation

Myasthenia Gravis — Tensilon® Test

Note ptosis

After the administration of tensilon

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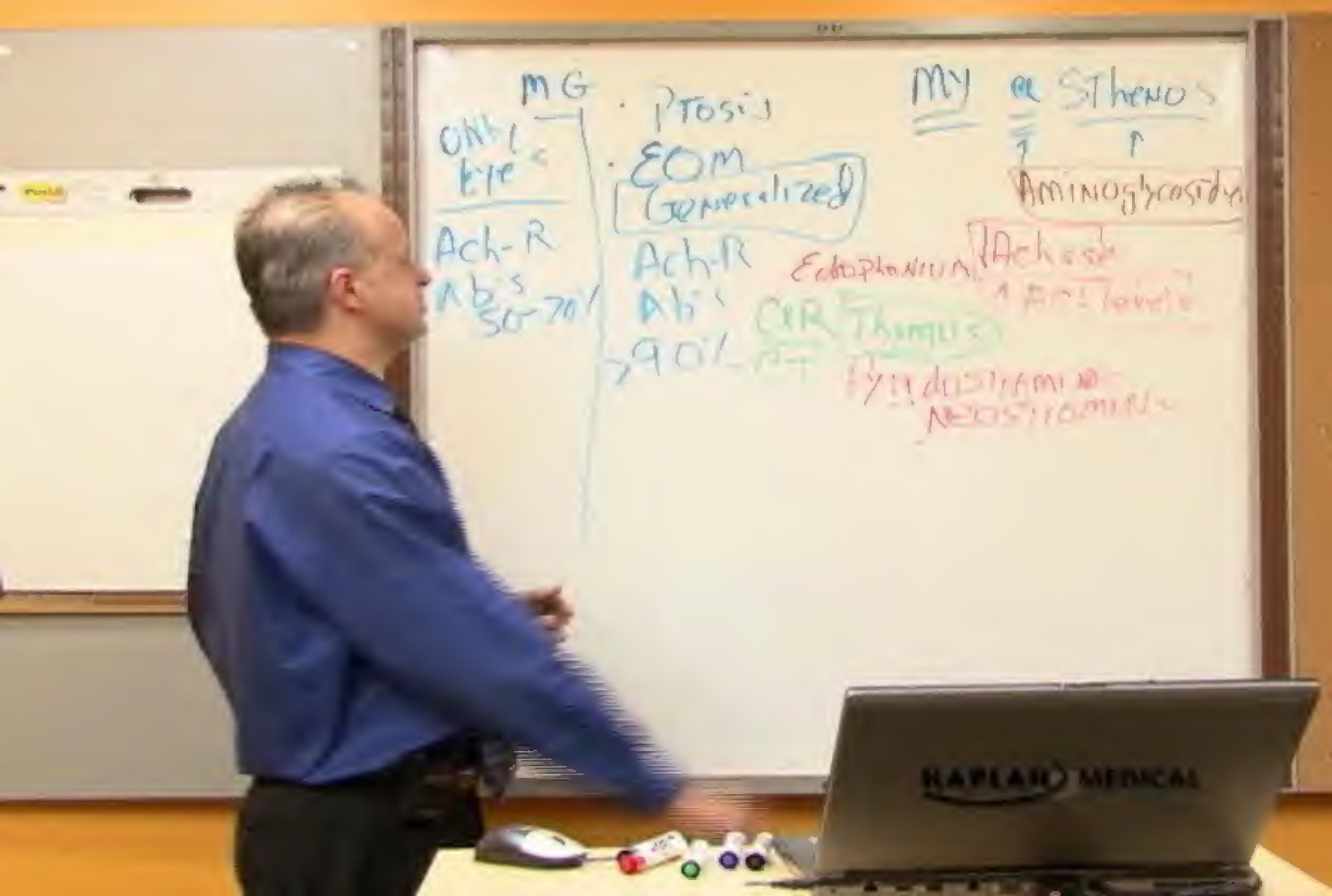
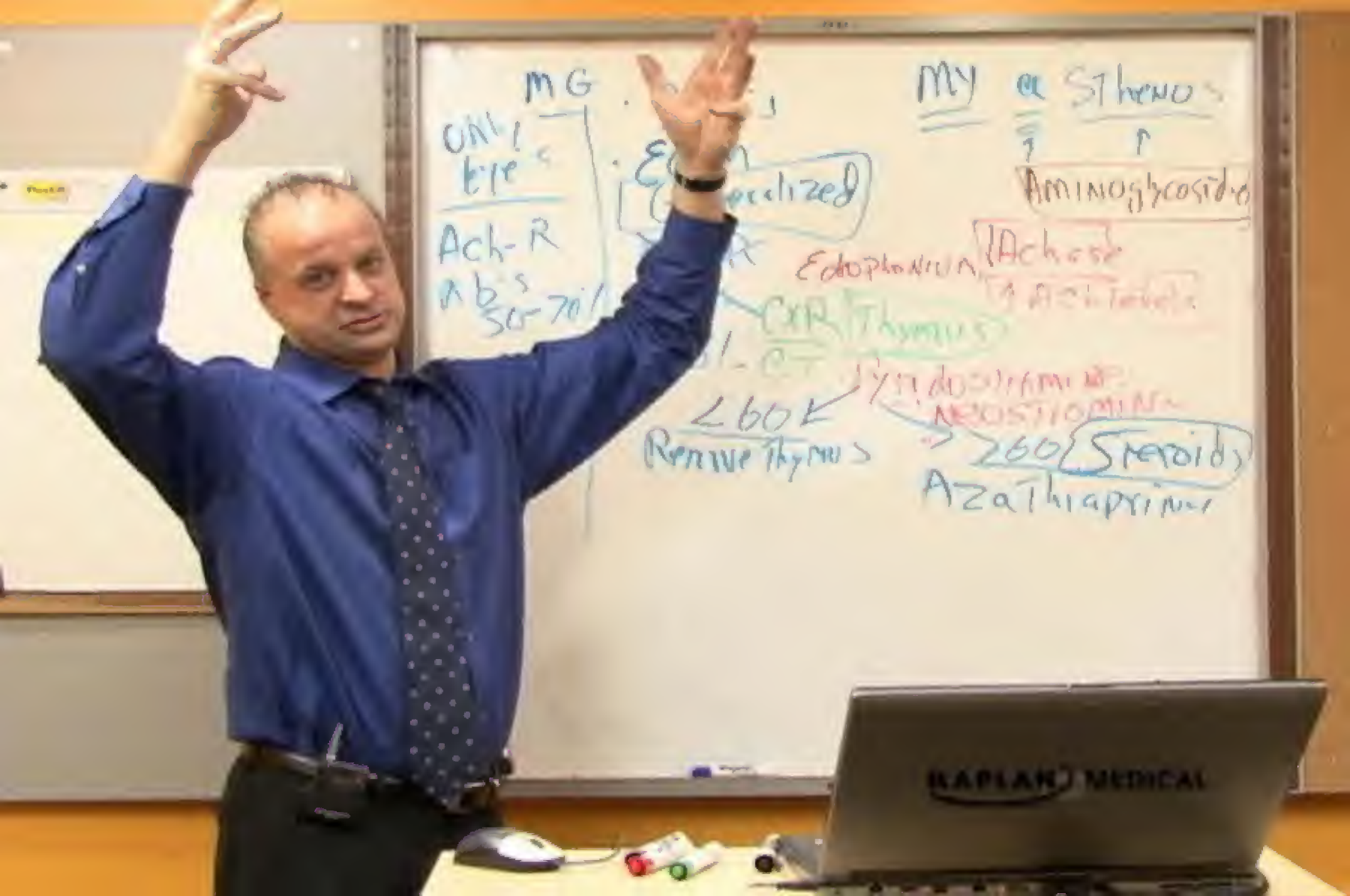
Myasthenia Gravis — Treatment

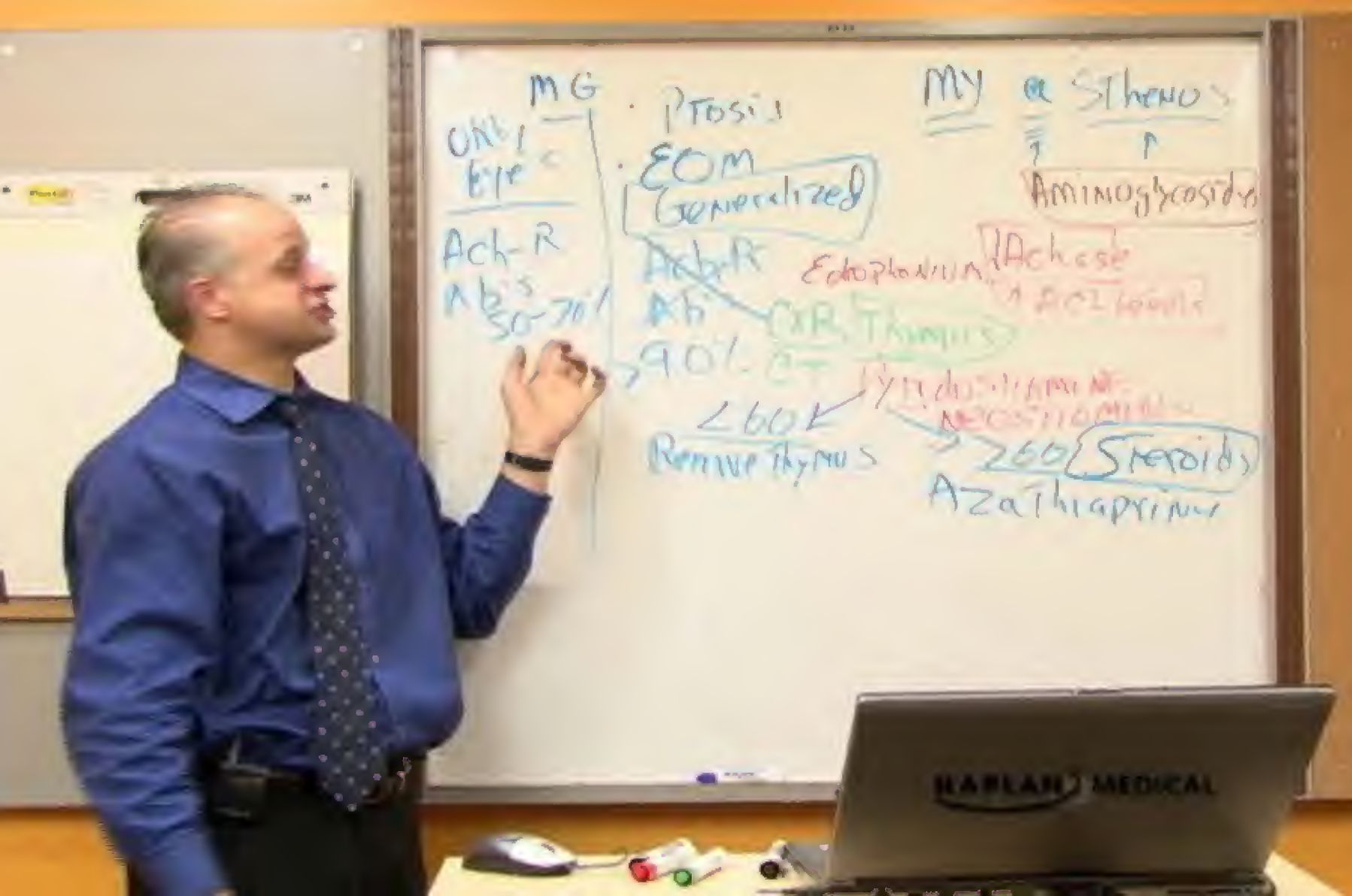
- Initial treatment for symptoms
  - Pyridostigmine or neostigmine
- If there is no response
  - Thymectomy
    - Post pubertal and < 60 years of age

Myasthenia Gravis — Treatment

- Still no response?
  - Immunosuppresive therapy
    - Steroids are the initial treatment of choice
    - If steroids fail- azathioprine usually added
    - Cyclosporine and cylophosphamide are alternatives (toxic)
    - If + respiratory symptoms: Plasmapheresis and intravenous immunoglobulin


MG
Ptosis
EOM
Generalized
Ach-R
Abs
MY
Asthenia
Aminoglycosides
Edrophonium
Thymus
Pyridostigmine
Neostigmine
HAPLARD MEDICAL


MG
MY
Ach-R
Aminoglycosides
Edrophonium
CXR
Thymus
260K
Remove thymus
260
Steroids
Azathioprine
KAPLAN MEDICAL


MG
Ptosis
EOM
Generalized
Ach-R
Ab's
50-70%
>90%
Edrophonium
CXR Thymus
<60
Remove thymus
>60
Steroids
Azathioprine
My
Asthenia
Aminoglycosides
HARLAND MEDICAL

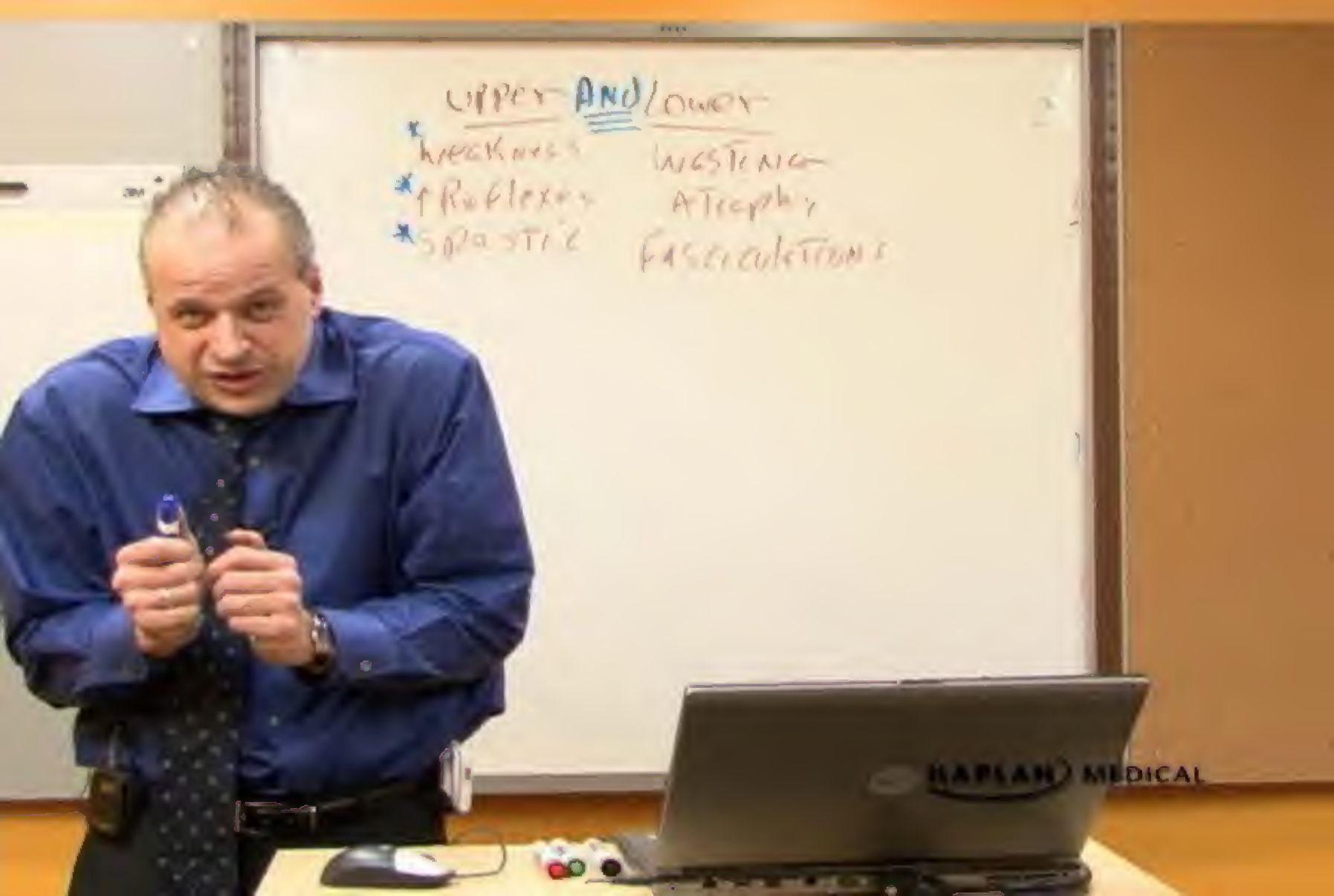
KAPLAN MEDICAL

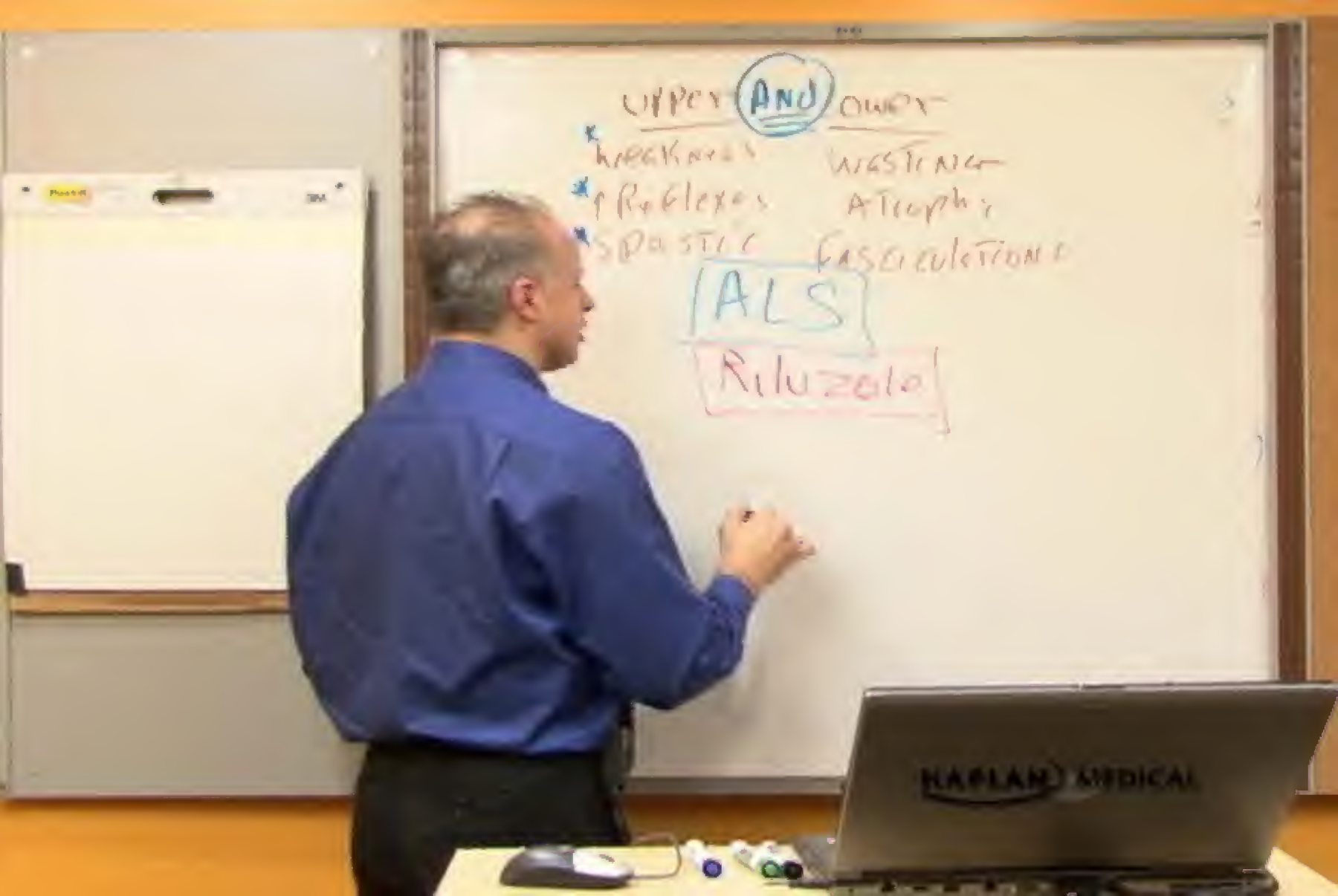
Ch. 6: Myasthenia Gravis

END

KAPLAN MEDICAL

Ch. 7: Amyotrophic Lateral Sclerosis (ALS)


UPPER AND/OR LOWER
* Weakness
* ↑ Reflexes
* Spastic
Weakness
Atrophy
Fasciculations
KAPLAN MEDICAL


AND
ALS
Riluzole
KAPLAN MEDICAL

KAPLAN

MEDICAL

Ch. 7: Amyotrophic Lateral Sclerosis (ALS)

END

KAPLAN MEDICAL

Ch. 8: Multiple Sclerosis

Multiple Sclerosis — Etiology

- Multifactorial
  - Genetic influence
  - Those above or below latitude 40 degrees North and 40 degrees South
    - Role for environmental trigger
      - » Infectious, dietary, climatic

Multiple Sclerosis — Clinical Presentation

Relapsing Remitting	Relapses of active disease with incomplete recovery between
Primary Progressive	Progressive from first onset with early disability
Secondary Progressive	Progressive with constant worsening of disease

Multiple Sclerosis — Diagnosis

- Clinical criteria
- Radiologic confirmation
  - MRI of the brain and spine most accurate
  - Increased T2 density and decreased T1 intensity in demyelinated plaques
  - Active MS lesions enhance with gadolinium and up to 6 weeks after

Multiple Sclerosis — Diagnosis

- Laboratory confirmation
  - CSF studies:
    - Mild pleocytosis, Mild elevation in total protein (levels >100 not MS)
  - If MRI of brain is negative, but suspicion is high
    - Check for oligoclonal bands in CSF

Multiple Sclerosis — Disease Modifying Therapy

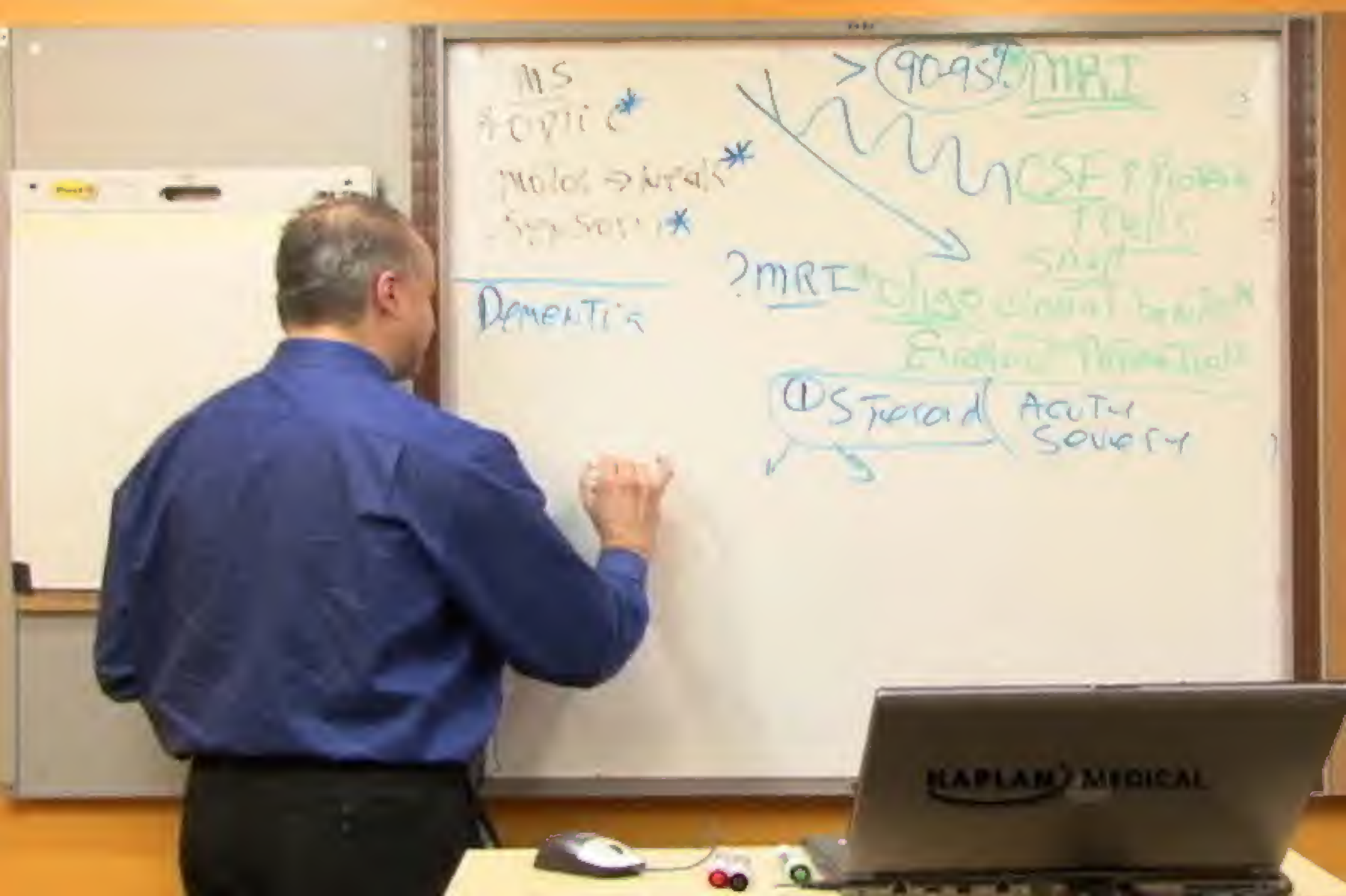
Relapsing Remitting	IFN-β1a IFN- β1b Glatiramer acetate (also known as copolymer I)
Primary Progressive	No approved therapy
Secondary Progressive	IFN- β1b Mitoxantrone

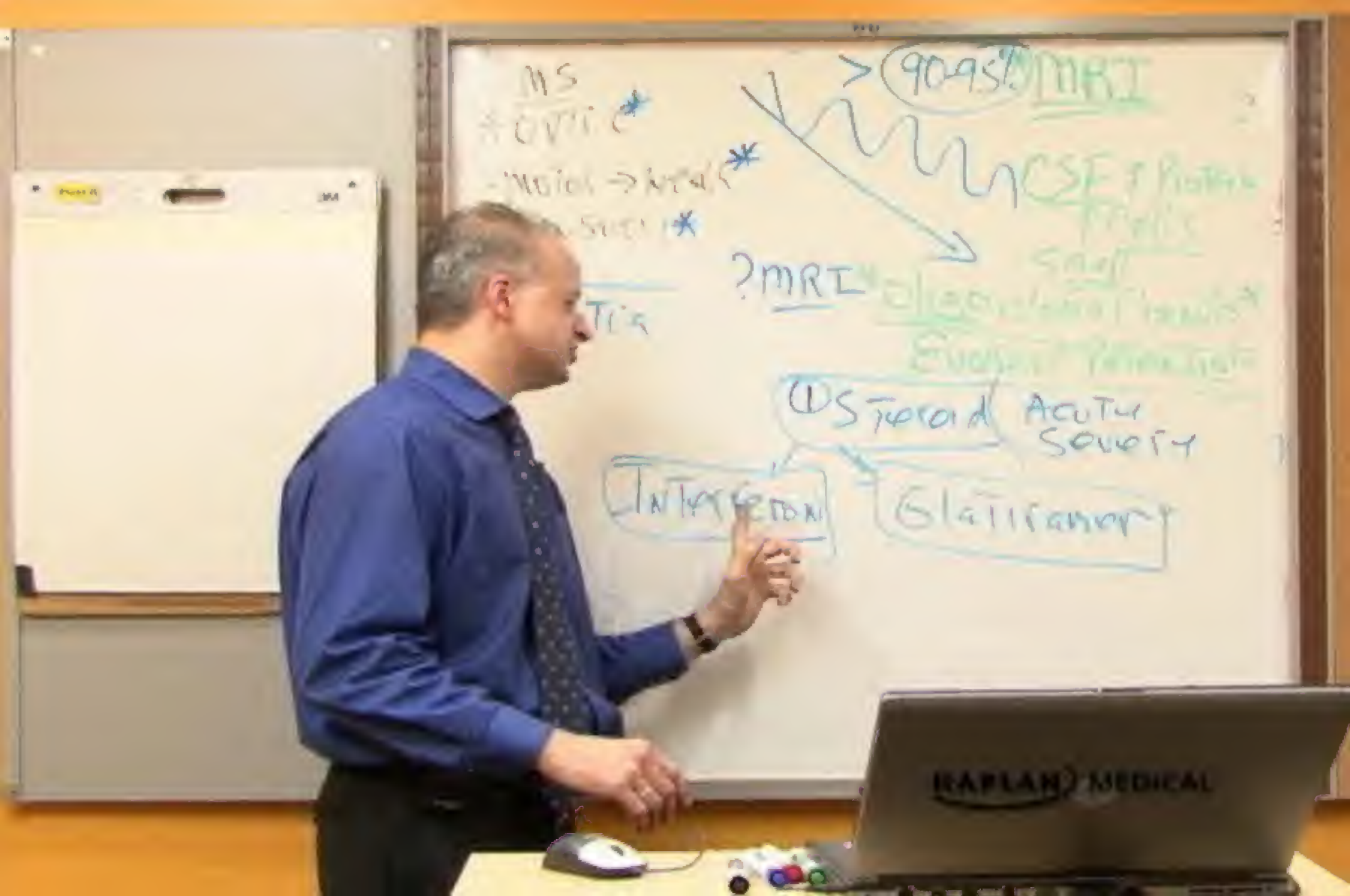
Multiple Sclerosis — Treatment of Acute Exacerbation

- Glucocorticoids x 3 days followed by 4 weeks per-oral taper
- Those unresponsive to steriods- plasma exchange

Multiple Sclerosis — Symptomatic Treatment

Spasticity	Baclofen (most effective) Tizandine and diazepam (nighttime)
Trigeminal neuralgia and dysesthesias	Carbamazepine, gabapentin, phenytoin, or TCAs
Bladder hyperreactivity	Oxybutynin
Urinary retention	Bethanechol
Fatigue	Amantidine or fluoxetine
Erectile dysfunction	Sildenafil


MRI
?MRI
DEMENTIA
ACUTE
KAPLAN MEDICAL


MS
>90-95%
MRI
?MRI
Steroid
Acute
Severity
Interferon
Glatiramer
RAPLAN MEDICAL

KAPLAN MEDICAL

Ch. 8: Multiple Sclerosis

END

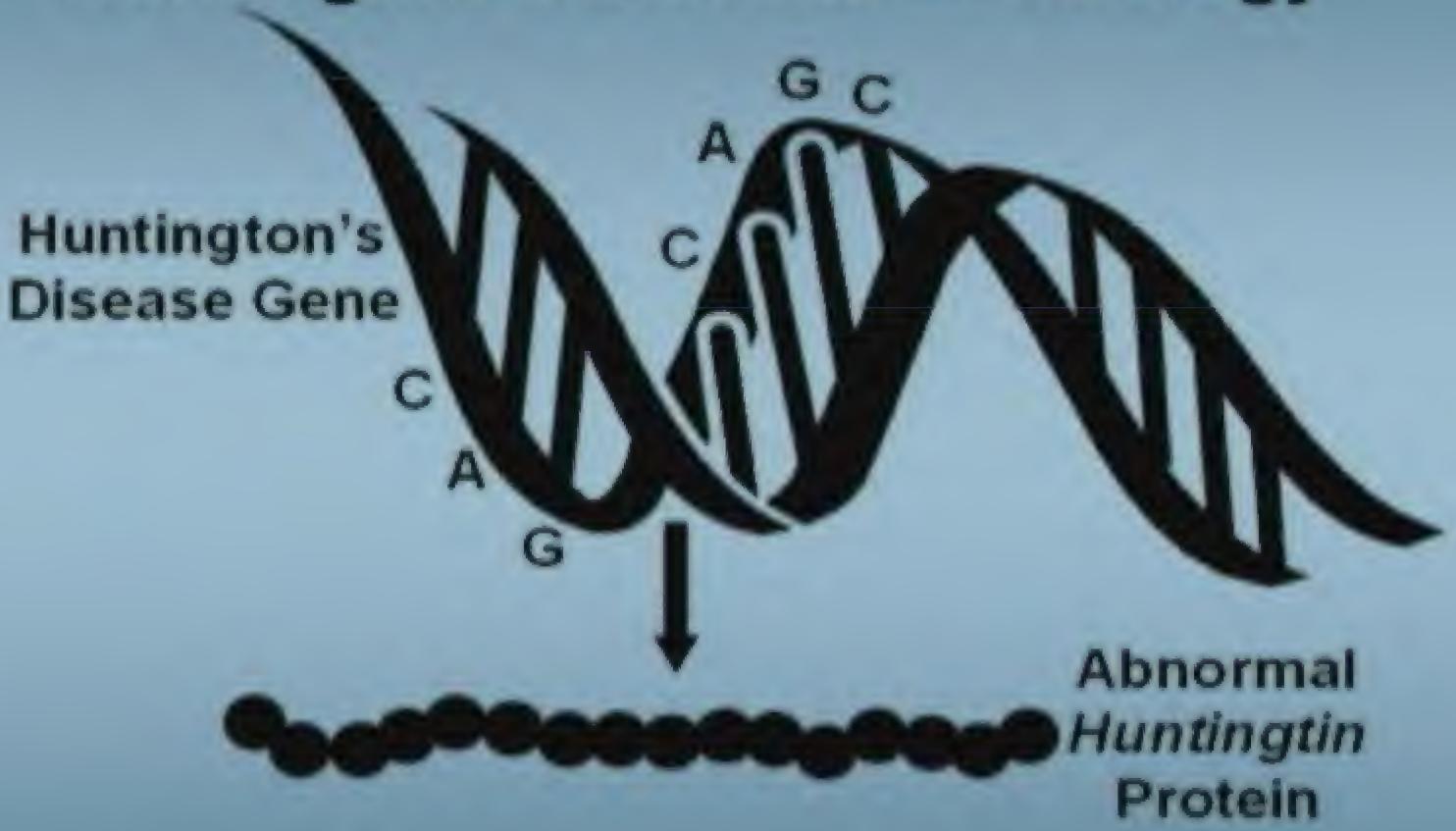
KAPLAN MEDICAL

Dementia: Huntington's Disease

Huntington's Disease — Etiology

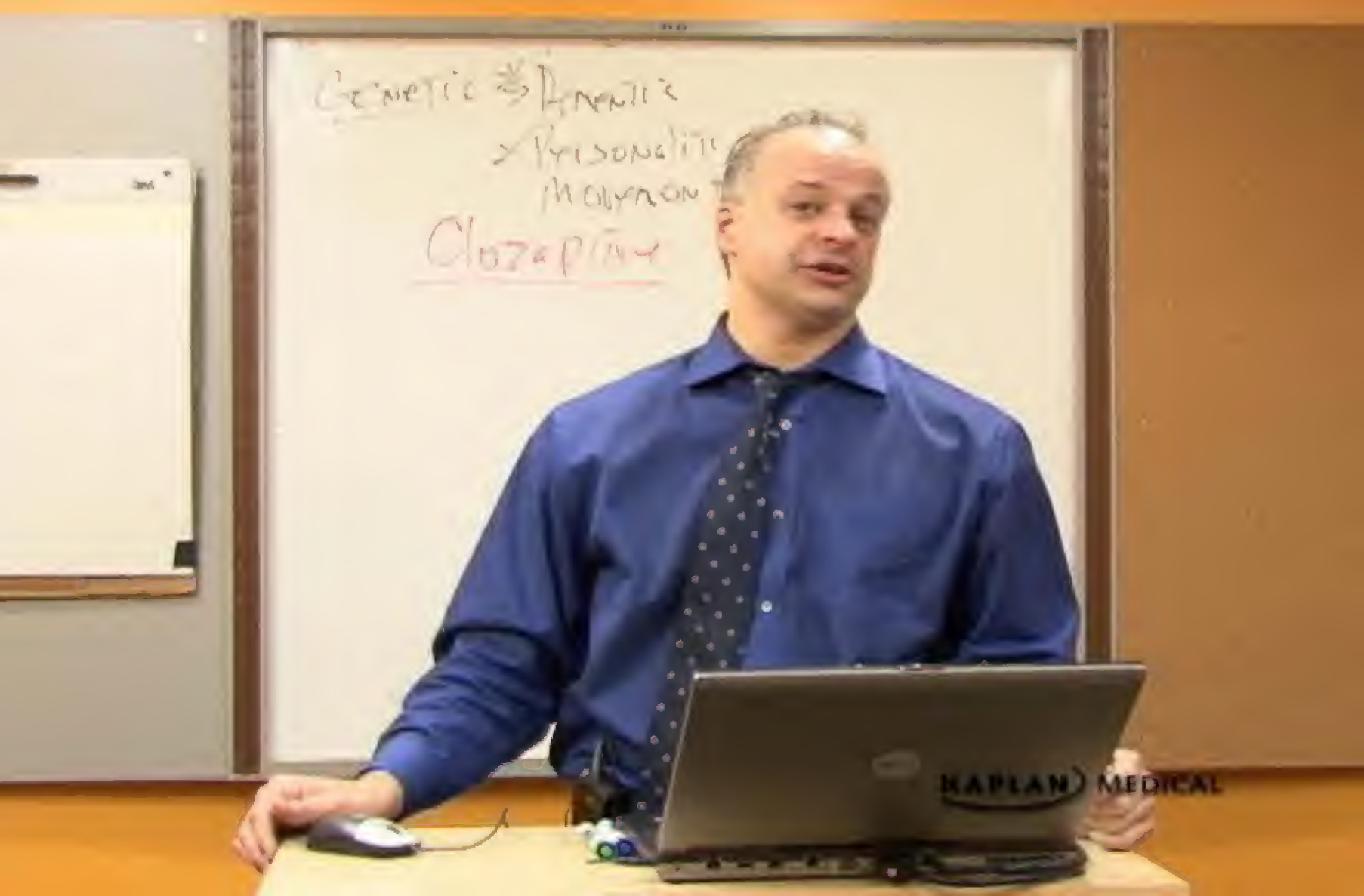
- Autosomal dominant
- Gene located on chromosome 4p codes for *Huntingtin* protein
- CAG trinucleotide repeat expansion
- Abnormal protein cleavage

Huntington's Disease — Etiology



Huntington's Disease — Diagnosis and Treatment

- Diagnosis
  - Genetic testing (DNA)
  - CT scans- cerebral atrophy, atrophy of the caudate late in disease
- Treatment
  - Clozapine for behavioral changes


Clozapine
KAPLAN MEDICAL

KAPLAN

MEDICAL

Dementia: Huntington's Disease

END

KAPLAN MEDICAL

Ch. 10: Parkinson's Disease and Other Movement Disorders

Parkinson's Disease — Syndromes

Supranuclear Palsy	Parkinsonism + vertical gaze palsy
Olivopontocerebellar Atrophy	Parkinsonism + prominent ataxia
Shy-Drager Syndrome	Parkinsonism + Prominent orthostatic hypotension

Parkinson's Disease — Treatment

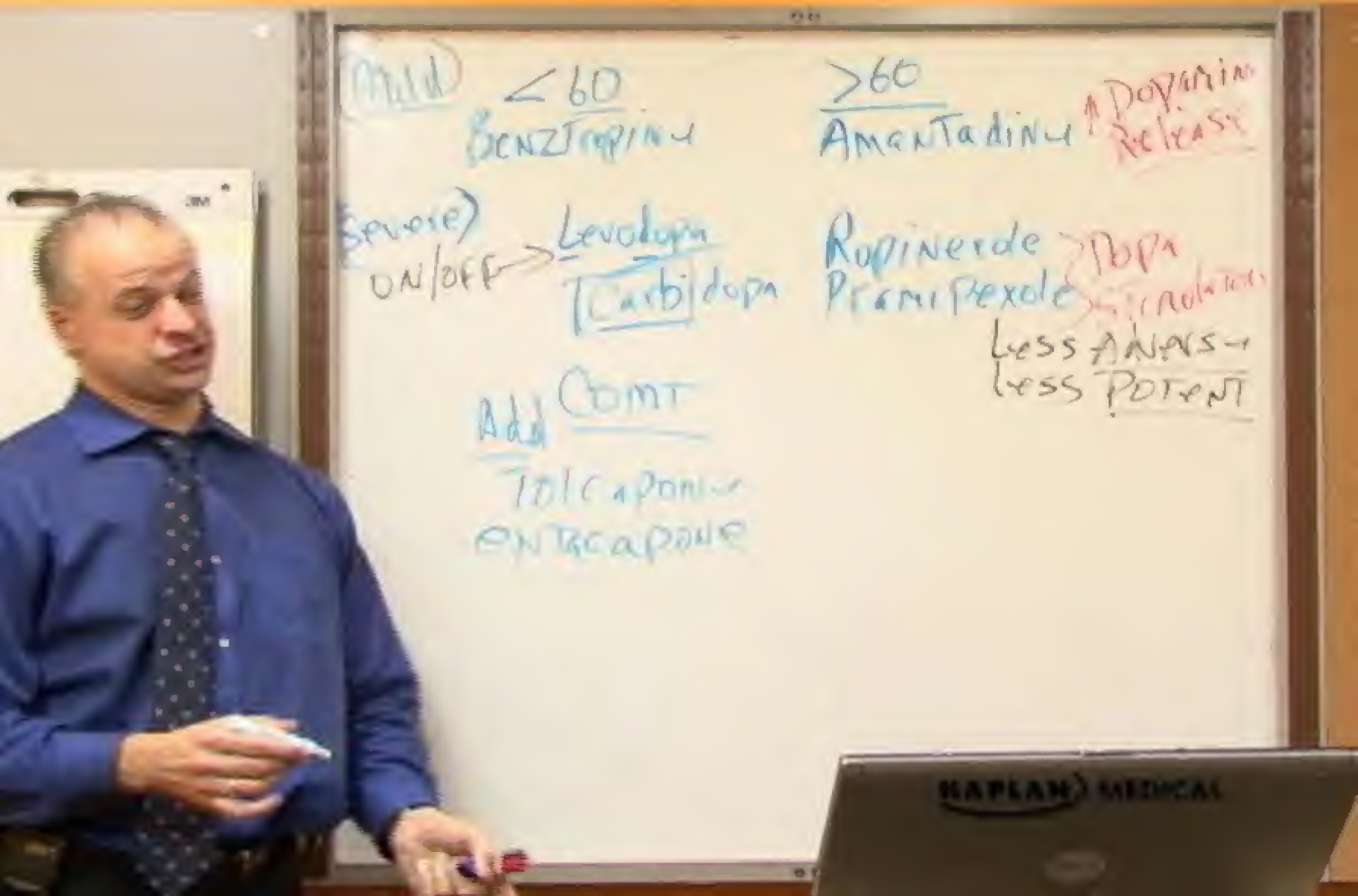
Dopamine agonists	Anticholinergics
• Carbidopa/ Levodopa •Pramiprexole, bromocriptine, pergolide and ropinirole (direct) •COMT inhibitors (tolcapone and entacapone), selegiline, amantadine (indirect)	• Benztropine • Trihexyphenidyl

Parkinson's Disease — Treatment

Patient Population	Drug of Choice
Functional status is intact and < 60 years of age	Start anti-cholinergic
Functional status is intact and > 60 years of age	Start amantidine
Compromised functional status	Start carbidopa/ levodopa


Orthostatic
KAPLAN MEDICAL


Orthostatic
Cerebellar
KAPLAN MEDICAL


Mild
< 60
Benztropine
> 60
Amantadine
↑ Dopamine Release
Severe
ON/OFF
Levodopa
Carbidopa
Ropinerole
Pramipexole
↑ Dopa
less Anes-
less Potent
Add
COMT
Tolcapone
Entacapone
KAPLAN MEDICAL

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MEDICAL

Parkinson's Disease

END

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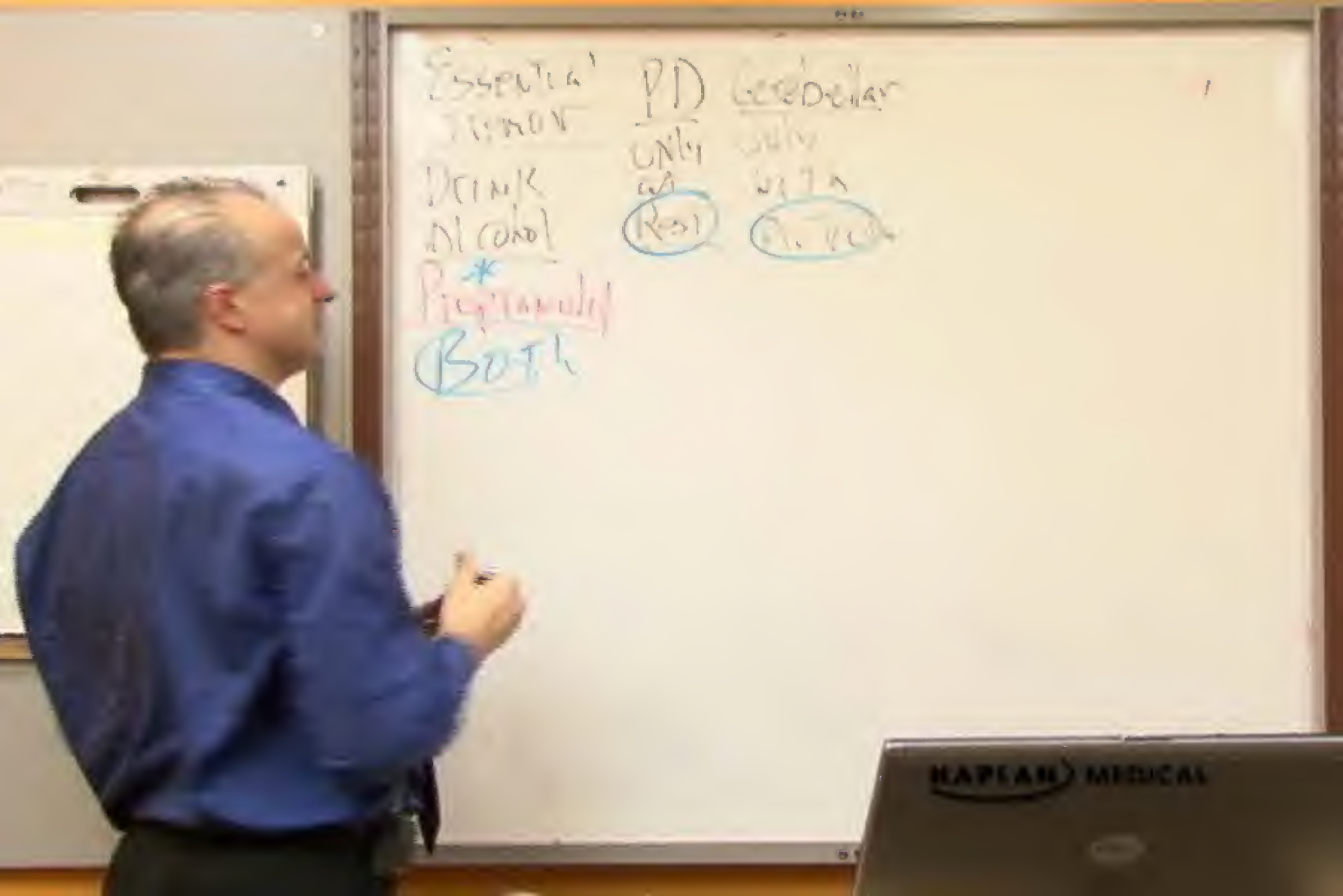
Parkinson's: Benign Essential Tremor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Parkinson's: Benign Essential Tremor

END

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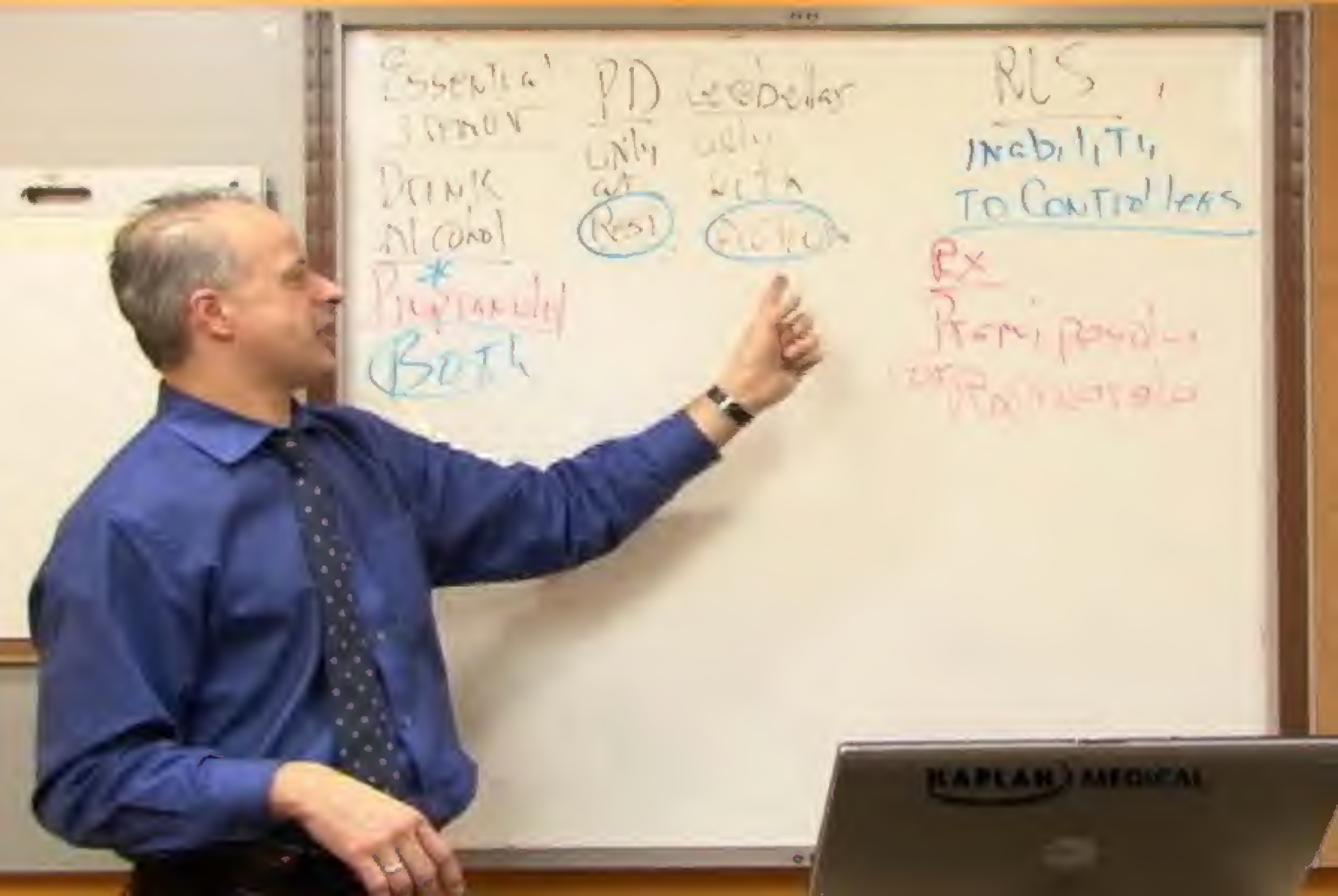
Parkinson's: Restless Leg Syndrome


Essential
Tremor
Drink
Alcohol
*
Primarily
Both
PD
Only
at
Rest
Cerebellar
only
with
Action
RLS
Inability
To Control Legs
Rx
Ropinirole
KAPLAN MEDICAL

KAPLAN

MEDICAL

Parkinson's: Restless Leg Syndrome

END